THEME 11 COGNITIVE AND PSYCHOLOGICAL ASSESSMENT AND SUPPORT

COG-01 Cognitive endophenotypes in ALS

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Background: Relatives of patients with amyotrophic lateral sclerosis (ALS) report a heightened rate of schizophrenia, suicide and other neuropsychiatric conditions (1). It is hypothesized that this is due to the pleiotropic (i.e. multiple) effect of risk genes such as C9orf72 (2).

Objective: This study aims to build on previous research by characterizing the cognitive profile of non-affected relatives of ALS patients.

Methods: One hundred and forty-nine first- and second-degree relatives of ALS patients and 60 controls completed a full neuropsychological assessment, including measures of intellectual functioning (Wechsler Abbreviated Scale of Intelligence- 2nd Edition; WASI-2), language (Boston Naming Test; BNT), executive functioning (Colour Word Interference Test; CWIT), Digit Span; DS, Verbal Fluency), social cognition (Reading the Mind in the Eyes Test; RMET) and memory (Rey Auditory Verbal Learning Test; RAVLT, Logical Memory from Wechsler Memory Scale – 3rd Edition; LM). ALS relatives and controls were compared on neuropsychological performance using Welch’s independent samples t-tests or Wilcoxon rank tests. Further sub-group comparisons using analysis of variance (ANOVA) were carried out to compare relatives of familial ALS patients (FALS; n = 91), relatives of sporadic ALS patients (SALS; n = 58), and controls; and comparing C9orf72 positive (n = 39) and negative ALS relatives (n = 158).

Results: ALS relatives scored significantly worse than controls on WASI-2 IQ (d = 0.54, 95% CI: 0.13, 0.95), phonemic verbal fluency (d = 0.81, 95% CI: 0.37, 1.24), CWIT inhibition errors (d = 0.49, 95% CI: 0.17, 0.80), DS backwards span (d = 0.8, 95% CI: 0.41, 1.19), BNT spontaneous correct (d = 0.91, 95% CI: 0.61, 1.20), RAVLT 5 trial total recall (d = 0.67, 95% CI: 0.26, 1.08) and LM delayed recall (d = 0.7, 95% CI: 0.29, 1.11). Post-hoc Bonferroni comparisons indicated that relatives of FALS patients showed particularly strong deficits in phonemic verbal fluency compared to controls (p < 0.001) and relatives of SALS patients (p < 0.05). Analysis of C9orf72 subgroups revealed few differences between C9orf72 positive and negative relatives, however, this is largely limited by the small numbers of C9orf72 positive relatives. Verbal fluency semantic score was significantly lower in C9orf72 negative than C9orf72 positive relatives (d = −1.25, 95% CI: −2.16, −0.31).

Conclusions: These findings suggest a cognitive endophenotype exists in non-affected members of ALS kindreds. Deficits on these tasks may signify disruption to underlying fronto-striatal networks in these individuals. These findings, if replicated, may improve statistical power in gene discovery studies, and lead to an improved understanding of the extended genetic, neuropsychological and neuropsychiatric profile in ALS.

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COG-02 ‘It refocuses the mind on the important things in life’: an exploration of burden and self-described positive experiences of informal ALS caregivers in Ireland and the Netherlands

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Background: Informal caregivers play a key role in the care of people with ALS. Research on caregiving generally focuses on the negative aspects including stress and burden (1). Consideration of positive aspects of caregiving is important also (2).

Objectives: To explore the complex self-reported burden and well-being experiences of informal caregivers at two leading ALS centres in Europe.

Methods: An exploratory cross-sectional study characterizes two groups of informal caregivers in Ireland (n = 76) and the Netherlands (n = 58). Data were collected as part of a European multi-centre study (3). In a semi-structured face-to-face interview, standardized measures assessed psychological distress, quality of life and burden, and in an open-ended question, caregivers were asked to identify any positive aspects of their caregiving experience. Descriptive statistics summarized the socio-demographic and wellbeing measures of the two cohorts. T-tests, Mann–Whitney U tests and Chi-square tests compared psychological distress, burden, and quality of life between caregiver cohorts. Thematic analysis was used to analyse the qualitative responses.

Results: Both caregiver cohorts were predominantly female and spouse/partners of the person with ALS. Higher levels of self-assessed burden were found among the Dutch caregivers (p = 0.0002), and higher levels of quality of life among the Irish cohort (p = 0.0004). Themes generated through qualitative analysis identified caregiver satisfaction, ability to meet the patient’s needs, the (re)evaluation of meaning and existential facets of life as positive aspects of caregiving. Existential factors were identified frequently by Irish caregivers (44%), and personal satisfaction and meeting the care recipient’s needs by the Dutch cohort (33.3% and 28.7%, respectively).

Discussion: It is important to generate an evidence base to support the psychosocial wellbeing and brain health of informal caregivers. Based on our findings, we suggest that both burden and the presence of positive factors should be evaluated and monitored. The possibility of concurrent positive and challenging experiences should be considered in the design and delivery of supportive interventions for informal caregivers. Further research will consider disease stage and quality of life between caregiver cohorts. The presence of positive factors should be evaluated and monitored. The possibility of concurrent positive and challenging experiences should be considered in the design and delivery of supportive interventions for informal caregivers.

Acknowledgements

Thank you to the caregivers who participated. Supported by funding from the Global Brain Health Institute, Alzheimer’s Association, and Alzheimer’s Society Pilot Awards for Global Brain Health Leaders (GBHI-ALZ-UK-20-638907). ALS-CarE study funding provided by the EU Joint Programme-Neurodegenerative Disease Research through the Health Research Board.

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COG-03 Theory of mind deficits in amyotrophic lateral sclerosis: a cross sectional population based study

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Objective: to assess Theory of Mind (ToM) abilities in a cohort of Amyotrophic Lateral Sclerosis (ALS) patients at diagnosis. Secondly, to assess possible differences in ToM abilities depending on cognitive profile, motor phenotype and degree of severity of motor symptoms.

Background: Affective and Cognitive ToM have been intensively studied in the last decades in several neurodegenerative diseases, including ALS (1). ToM deficits have been included in ALS-FTD revised diagnostic criteria (2). There is, however, remarkable variability in ToM impairment profile across different studies and this may be related to the high cognitive and motor phenotypic heterogeneity in ALS patients and variability in Neuropsychological tools employed (3).

Methods: We included 85 patients attending the ALS Centre of Turin University Hospital between May 2019 and June 2021. All patients underwent neurological evaluation and cognitive evaluation. Affective ToM assessment was performed through Reading Mind in the Eyes task (RMET) and Story-Based Empathy task-Emotion Attribution (SET-EA), while cognitive ToM assessment was performed through Story-Based Empathy task-Intention Attribution (SET-IA). Both SET-IA and SET-EA were compared to a control condition of Causal Inference (SET-C).

Results: Out of 85 patients included, 4 (5%) were diagnosed with ALS-FTD, 9 (11%) ALScbi, 23 (27%) ALSbi, 5 (6%) ALSbi and 44 (52%) were cognitively normal (ALS-CN), according to the revised ALS-FTD Consensus Criteria. Overall, 15 patients (18%) showed deficit in at least one of ToM tests. Specifically, RMET corrected scores showed a positive significant correlation with Category Fluency test (p = 0.02), while SET-EA and SET-IA corrected score showed significant correlation with Trial Making Test B-A (p = 0.02 and 0.006, respectively). Moreover, RMET showed significant correlation with onset site (p = 0.004, with bulbar onset patients performing worse than spinal onset ones), and degree of severity of motor symptoms expressed through ALSFRS-R (p = 0.009).

Conclusions: Deficits in affective and cognitive ToM may be present in ALS patients since the early stages of the disease and show a significant correlation with degree of severity of motor symptoms and executive functions. The presence of ToM deficits in ALS patients since the initial disease stages suggests the need of a systematic assessment, both to provide a more accurate definition of cognitive profile across FTD-ALS spectrum, and also to help clinicians to elaborate better communication and care strategies taking into account such deficits in ALS patients.
Cognitive and psychological assessment and support

Acknowledgements

Authors thank all the participant involved in the study. Authors declare that this abstract is original, has not been published before and is not currently being considered for publication elsewhere. The authors declare that there is no conflict of interest.

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COG-04 A scoping review of the literature to inform psychological support interventions for informal caregivers of people with motor neuron disease

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Background: Motor neuron disease (MND) is a group of terminal, neurodegenerative diseases that cause progressive, and often rapid, paralysis. Unsurprisingly, diagnosis and disease progression are also very distressing for family members and friends, who often become informal carers, and studies of the experiences of MND carers often highlight the need for psychological support. Therefore, this project aims to explore psychological interventions that could be beneficial for informal carers of people living with MND to help inform further research and service improvements.

Method: A scoping review was carried out to explore and describe psychological interventions for MND carers. Initially, search terms included MND, synonyms for MND, and variants of MND, as well as search terms for carers (including synonyms and terms for different relatives, such as ‘family’, ‘spouse’, ‘partner’, ‘daughter’, ‘son’, ‘friend’, etc.), and terms for psychological interventions. The goal was to be as inclusive as possible. Given the small amount of research available specifically focused on MND carers, the searches were then expanded to also include studies of psychological interventions for carers of people with related diseases, whose needs might partially overlap. This included carers of people with Parkinson’s disease, Huntington’s disease, and multiple sclerosis, and the search terms were expanded accordingly. The literature was collated from the databases: MEDLINE via OvidSP and PsycInfo (APA PsycInfo5) via Ovid and subsequently screened for relevance.

Results: Interventions were generally delivered face-to-face; the majority of interventions were targeted towards current caregivers aged 18 years old or above. Outcome measures generally focused on anxiety and depression. Results of the studies were often inconclusive due to small numbers of participants, lack of control conditions, and high attrition rates. However, there were some positive results for interventions incorporating mindfulness and meditation. In addition, across all of the intervention types, participant feedback was positive, with many caregivers saying they would recommend the support to others and reporting that they valued the peer support offered by group interventions.

Conclusion: This scoping review provides a useful starting point for future research to develop/adapt psychological interventions for MND carers. Carers who took part in the interventions generally provided positive feedback, and interventions including mindfulness and meditation show some promise. Future research should build on these findings, with a particular focus on refining interventions to support MND carers, and increasing accessibility to ensure more carers can fully participate in the interventions.

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COG-05 Application of executive function composite score (UDS3-EF) in a cohort of ALS-FTD spectrum patients

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Background: As many as 50% of ALS patients experience cognitive and/or behavioral symptoms, with approximately 15% developing dementia. Conversely, approximately 15% of FTD patients develop motor symptoms. Despite the growing understanding of this overlap between these two disorders, there remains a gap in readily assessing and uniformly quantifying cognitive dysfunction in this group of patients.

Objectives: We aimed to utilize the Uniform Data Set (v3.0) executive function composite score (UDS3-EF) model (1) in an NIH ALS-FTD disease spectrum cohort to uniformly assess executive function. The UDS validation cohort included clinically normal, MCI, AD, and bvFTD patients. However, it did not include ALS patients or patients with other diagnoses in the ALS-FTD spectrum.

Methods: Our cohort (n = 78) is 52.6% female, 93.6% white, and 16.2 average years of education and includes 24 ALS and 36 FTD patients as well as 18 patients diagnosed with other neurodegenerative conditions. Because the demographics of the NIH cohort were well-matched to that of the cohort analyzed to develop the UDS model, we were able to utilize the demographics-adjusted model for EF scores. Parametric and non-parametric statistical tests were chosen based on variable distribution.

Results: The median EF scores for ALS (−0.491; IQR −1.466, −0.012) were significantly higher than that of FTD (−2.868; IQR −3.697, −1.708); Mann–Whitney U = 94, p < 0.0001, representing greater executive function. In 75% of ALS patients,
the EF scores were under zero indicating executive function impairment. Patients across the full FTD spectrum, beyond bvFTD (including patients diagnosed with CBS, PPA, and PSP), had abnormal scores consistent with impaired executive function.

**Conclusion/Discussion:** The UDS3-EF model may be used in ALS-FTD spectrum patients to allow for greater insight into the severity of executive dysfunction in these patients. These results indicate that a high proportion of ALS patients have impaired executive function similar to previous reports, and as expected, more severe impairment is present across the FTD disease spectrum. These results have important implications for recognizing impaired cognition in this group of patients, which impacts their daily lives, from self-care to management of finances and beyond.

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**COG-06 Plasma uric acid helps predict cognitive impairment in patients with amyotrophic lateral sclerosis**

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**Objective:** To identify potential clinical and biochemical biomarkers of cognitive impairment in patients with amyotrophic lateral sclerosis (ALS), we examined the differences in clinical and biochemical parameters between patients with cognitive impairment (ALS-ci) and those with no cognitive impairment (ALS-nci), and evaluated the predictive power of potential parameters.

**Methods:** In this cross-sectional study, 124 ALS patients were assessed by the Edinburgh cognitive and behavioral screen (ECAS) (¹) and categorized into the ALS-ci group and the ALS-nci group. Clinical, biochemical, and behavioral data were compared between the two groups. Parameters with significant difference were further included in a multivariate logistic regression analysis and assessed for their predictive power of cognitive impairment in ALS patients.

**Results:** Up to 40% of ALS patients showed cognitive impairment. The ALS-ci group had lower education level (p < 0.001), older age at symptom onset (p = 0.001), and older age at testing (p = 0.001). Multivariable analysis showed that low plasma uric acid (p = 0.037), older age at testing (p = 0.002) and low education level (p = 0.001) were independent predictors of cognitive impairment in ALS patients. The predictive model including these three parameters had an area under the curve value of 0.788 with a sensitivity of 79.2% and a specificity of 73.7%.

**Conclusion:** Cognitive impairment was a relatively common feature in ALS patients. Plasma uric acid might help evaluate the risk of cognitive impairment in ALS patients when combined with education level and age at testing.

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**Acknowledgements**

The authors thank all patients, caregivers, and doctors from the Department of Neurology, Tongji Hospital for their support. This work was funded by the clinical research program of Bethune Charitable Foundation.

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**COG-07 Brainstem correlates of pathological laughter and crying frequency in ALS**

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Pseudobulbar affect is a disorder of emotional expression commonly observed in amyotrophic lateral sclerosis (ALS), presenting as episodes of involuntary laughter, or crying. The objective of the current study was to determine the association between the frequency of pathological laughter and crying (PLC) episodes with clinical features, cognitive impairment, and brainstem pathology. 35 sporadic ALS patients underwent neuropsychological assessment, with a subset also undergoing brain imaging. The Centre for Neurological Study Lability Scale (CNS-LS) was used to screen for the presence and severity of pseudobulbar affect (CNS-LS ≥ 13) and frequency of PLC episodes. Presence of pseudobulbar affect was significantly higher in bulbar onset ALS (p = 0.02). Frequency of PLC episodes was differentially associated with cognitive performance and brainstem integrity. Notably pathological laughter frequency, but not crying, showed a significant positive association with executive dysfunction on the Trail Making Test B-A (R² = 0.14, p = 0.04). Similarly, only pathological laughter frequency demonstrated a significant negative correlation with grey matter volume of the brainstem (R² = 0.46, p < 0.01), and mean fractional anisotropy of the superior cerebellar peduncles (left: R² = 0.44, p < 0.01; right: R² = 0.44, p < 0.01). Hierarchical regression indicated brainstem imaging in combination with site of symptom onset explained 73% of the variance in pathological laughter frequency in ALS. The current findings suggest emotional lability is underpinned by degeneration across distinct
neural circuits, with brainstem integrity critical in the emergence of pathological laughter.

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COG-08 Vascular risk factors decrease the risk of cognitive impairment in amyotrophic lateral sclerosis: a case-control study

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Background: The disease-modifying effects of diabetes mellitus (DM) (1), hyperlipidemia (2), and overweight (3) on risk and prognosis of ALS have gained significant attention in recent years. However, whether these well-known vascular risk factors increase the cognitive burden in patients with ALS remains unclear.

Objectives: To evaluate the association between vascular risk factors (including hypertension, DM, hyperlipidemia, overweight and smoking) and cognitive function in patients with ALS.

Methods: Patients with ALS were consecutively recruited between June 2012 and November 2019 from a tertiary referral center for ALS at the West China Hospital. Vascular risk factors were confirmed based on clinical data, while cognitive function was evaluated by the Chinese version of the Addenbrooke's Cognitive Examination-revised. Case-control design to investigate the association between vascular risk factors and cognitive impairment in ALS. With careful confounder adjustment, multivariable logistic regression analysis was performed separately (for each factor) and accumulatively (based on the sum of factors) to determine the association between cognitive impairment and vascular risk factors in ALS.

Results: Of 870 patients, 561 (64.5%) were men, the mean age at registration was 54.1 (11.3) years and 266 had cognitive impairment. No cognitive burden from vascular risk factors was found in patients with ALS. On the contrary, we first observed that DM (odds ratio [OR], 0.50; 95% confidence interval [CI], 0.25–0.98; p = 0.04) and hyperlipidemia (OR, 0.50; 95% CI, 0.26–0.97; p = 0.04) showed protective effects against cognitive decline in ALS, adjusted for age, sex, educational level, site of onset, Revised-ALS Functional Rating Scale score, predominant upper motor neuron phenotype, family history of ALS, and the remaining vascular risk factors. Furthermore, patients with >2 vascular risk factors showed a significantly lower risk of cognitive impairment (OR, 0.18; 95% CI, 0.07–0.48; p = 0.001). Sensitivity analyses of sex did not substantially reverse the risk estimates.

Discussion: This study suggests that vascular risk factors, particularly DM and hyperlipidemia are associated with cognitive benefits in patients with ALS. This neuroprotective effect adds new evidence to the fitness hypothesis and highlights the unique aspects of ALS, both in clinical heterogeneity and pathophysiological complexity. Elucidation of mechanisms behind these observed associations might open new perspectives in ALS therapeutics.

Acknowledgements
We would like to thank the patients and their families for taking part in the study.

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COG-09 Exploring the psychological support preferences of informal carers of people with motor neuron disease

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Background: Given the nature of MND, many family members and friends become informal carers, giving up their own time, including their own professional, personal, and social lives. Previous research has highlighted the unmet needs of MND informal carers; however, there remains a gap in our understanding of the needs (and how to address said needs) specific to MND carers.

Objectives: This study aims to explore the needs and support preferences of informal MND carers in more detail, including directly through 1:1 discussion, thereby informing the future development of a psychological intervention.

Methods: Previously, we have collected mood scores of depression (PHQ-9), anxiety (GAD-7), and strain (MCSI) from informal MND carers using the Telehealth in MND (TiM) system. In the present study, this data was analysed using descriptive statistics. New participants, consisting of both current and bereaved informal carers, were recruited via an advert circulated on social media and disseminated by the MND Association. Carer experiences, challenges, unmet needs, and preferences for psychological support were explored further in semi-structured interviews, which were held remotely and transcribed. Thematic analysis from a grounded-inductive perspective was used to identify common themes and key elements, which may serve as therapeutic targets.

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Results: Mood scores were collected from 35 caregiver participants across multiple time points; measures included the PHQ-9 and GAD-7. Thirty-one percent of participants (n = 11) reported a clinical level of depression on at least one occasion, while forty percent (n = 14) reported a clinical level of anxiety at least once. MCSI scores suggest a high level of caregiver burden. Thematic analysis is ongoing, but emerging observations include: participants almost unanimously in favour of a remote (video call) format for psychological support sessions; several suggestions for an ‘emergency hotline’ type of telephone support, either by a professional or knowledgeable volunteer; reports of feeling emotionally compelled to support the relative with MND, with some participants expressing discomfort at leaving their family member under formal care despite the strain their informal role creates; and most participants feeling inadequately supported by MDT members and healthcare professionals due to a lack of empathy, specialist knowledge, and/or training.

Discussion: We have identified evidence of mood disturbance among informal MND carers; experiences of depression and anxiety fluctuated over time suggesting these factors may be ameliorable to an intervention. We identified additional areas of psychological distress, barriers to accessing support, and preferences for the type of support they require. The outcome of this pilot study will help to inform the development of a new psychological intervention tailored to informal carers of MND.

COG-10 ‘Seeing my wife gradually deteriorate and knowing where it is going makes me sad’: a multicentre, exploratory study of burden and difficulties of informal ALS caregivers

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Background: It is essential to support the wellbeing of Informal caregivers, as they play a vital role in caring for the person with ALS (1).

Objectives: To explore the burden and difficulties of caring for someone with ALS in three leading ALS centres in Europe.

Methods: This mixed-methods analysis describes the burden and self-defined difficulties of informal ALS caregivers from three multidisciplinary ALS centres – Dublin, Ireland (n = 76), Utrecht, the Netherlands (n = 58) and Sheffield, England (n = 38). Data were collected as part of a European multicentre study (2). In face-to-face semi-structured interviews standardised measures assessed burden, quality of life and psychological distress; an open-ended question asked about difficult aspects of caregiving. Descriptive statistics summarized socio-demographic and wellbeing measures, and statistical tests were used to explore the characteristics and wellbeing measures across cohorts. Thematic analysis was used to analyse qualitative responses.

Results: Most caregivers were female, spouses/partners, and lived with the person with ALS. Irish caregivers had significantly lower levels of burden than the Dutch and English cohorts (p = .001). The Dutch cohort had significantly lower levels of anxiety (p = 0.016) and quality of life (p = 0.012) than the Irish cohort. For quality of life and anxiety, the English cohort fell between the other two. There were no significant differences between cohorts for depression. When asked to detail difficulties associated with caregiving, carers described the practical issues associated with the caregiver role, restrictions on lives and livelihoods, and the impact on their health. Psycho-emotional factors identified included stress, worry, witnessing patient decline and relationship changes. The frequency of themes varied across the three cohorts.

Discussion: The psychological composition and subjective experience of caregivers are important factors influencing burden (3). This mixed-methods approach allows a nuanced understanding of burden and difficulties experienced. We offer a description of caregivers at three ALS centres and provide a strong evidence base for more detailed analysis of caregiver burden experiences. Further research will consider disease stage, patient cognitive/behavioural status and the influence of cultural context on caregiver wellbeing.

Acknowledgements

Thank you to the caregivers who participated in this research. This work was supported by funding from the Global Brain Health Institute, Alzheimer’s Association, and Alzheimer’s Society Pilot Awards for Global Brain Health Leaders (GBHI ALZ UK-20-638907). The ALS-CarE study was funded by the EU Joint Programme – Neurodegenerative Disease Research through the Health R

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COG-11 Exploring potential markers of pre-dementia risk states in motor neuron diseases: a longitudinal study of mild behavioral impairment and its relation to cognition

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Background: Mild behavioral impairment (MBI) is a neurobehavioral syndrome characterized by later-life emergence of sustained neuropsychiatric symptoms, and represents the neurobehavioral axis of pre-dementia risk states as a complement to the neurocognitive risk axis represented by mild cognitive impairment. Overt behavioral symptoms have been frequently reported in motor neuron diseases (MNDs), but a specific investigation of MBI as a potential marker of pre-dementia risk states is still lacking.

Objectives: To evaluate MBI in MNDs both at onset and over disease course, and to explore its relationship with baseline and longitudinal cognitive features.

Methods: 56 MND patients with complete cognitive/behavioral and mood examinations were included. Patients were followed longitudinally with mood and behavioral evaluations approximately every 3 months, and with cognitive examinations approximately every 6 months, for up to 15 months. Cognitive/behavioral alterations were assessed using the Edinburgh Cognitive and Behavioural ALS Screen (ECAS). MBI was evaluated using the MBI checklist (MBI-C) and mood disturbances were assessed using the Hospital Anxiety and Depression Scale (HADS). Associations between baseline MBI-C domains scores and baseline cognitive and mood symptoms were tested using the Spearman's correlation coefficient. Based on longitudinal data individual slopes of decline for each cognitive measure were generated, and linear regression models were used to evaluate the role of baseline MBI-C domains scores in predicting longitudinal rates of cognitive decline.

Results: At baseline 30.35% of patients manifested MBI, and diagnoses of MND with cognitive and/or behavioral impairment (MND-CBI) were significantly more frequent in patients with MBI (p < 0.001). The most impaired MBI domain was affective/ emotional dysregulation, followed by impulse dyscontrol, apathy, social inappropriateness, and abnormal thoughts/perception. Apathy was significantly associated with worse general cognition (lower ECAS total scores, p = 0.04) and more impaired ALS non-specific functions (lower ECAS visuospatial scores, p = 0.04), affective/emotional dysregulation was significantly associated with greater depressive symptoms (higher HADS depression scores, p = 0.02), and impulse dyscontrol was significantly associated with more impaired ALS specific functions (lower ECAS ALS specific scores p = 0.03, ECAS executive scores p = 0.05 and ECAS language scores p = 0.04). 60.71% of patients underwent at least one longitudinal examination and, among them, 14.70% developed MBI. The greatest longitudinal decline was observed in the affective/emotional dysregulation domain, followed by apathy, impulse dyscontrol and social inappropriateness domains. Greater baseline scores in the impulse dyscontrol and abnormal thoughts/perception domains were significant predictors of a greater decline in the executive (p < 0.001 and p = 0.007) and fluency domains (p = 0.001 and p < 0.001). Greater baseline scores in the apathy domain were significant predictors of a greater decline in the visuospatial domain (p = 0.04).

Discussion: Our results suggest that MBI might represent an early marker of incident cognitive decline in patients with MND, and provide preliminary evidences supporting the usefulness of MBI evaluation to detect dementia in its preclinical/prodromal phase.

COG-12 Subjective disease burden due to cognitive and behavioural impairment in amyotrophic lateral sclerosis compared to frontotemporal dementia and Parkinson’s disease

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Background: The influence of cognitive and behavioural changes on subjective burden (sTBS) in ALS, idiopathic Parkinson’s disease (IPD) and frontotemporal dementia (FTD) have never been compared before in a study.

Objectives: This cross-sectional study investigates the disease burden of ALS, IPD and FTD patients by evaluating the QoL and affective state as measures of psychosocial adaptation to cognitive and behavioural impairments, while taking disease insight into account.

Methods: In a prospective design, cognitive and behavioral impairments of N = 40 ALS, N = 40 IPD and N = 17 FTD patients and the ratings of N = 31 ALS-, N = 30 IPD- and N = 16 FTD-caregivers were assessed to compare disease burden. Cognition was measured with the Edinburgh Cognitive and Behavioral Screen (ECAS) and Mini Mental State Examination (MMSE). Behavioural impairments were estimated by a caregiver with the ECAS behavioural questionnaire. The disease burden questionnaire had a self-rated and a caregiver-estimated version for the patients’ burden. The discrepancy of both scores was calculated to evaluate the degree of disease insight. Quality of life (QoL) was assessed with the schedule for the evaluation of individual QoL, anamnestic comparative self-assessment and Health related Quality of life (HRQoL) with Short Form 12. Affect was measured with ALS Depression Inventory 12. Physical impairment was assessed by ALS Functional Rating Scale (ALS-FRS), Unified Parkinson’s Disease Rating Scale [UPDRS] and Hoehn & Yahr [H&Y].

Results: All three groups maintained a comparable, satisfactory QoL and good affective state. The ALS group had the
lowest physical HRQoL but still a good mental QoL, followed by the IPS group. Cognitive and behavioral impairments, depressive symptoms significantly positively correlated with a higher sTBS in IPS and ALS groups. Additionally, in the IPS group, physical impairment and longer disease duration significantly correlated with a higher sTBS and lower QoL. The sTBS in the FTD group was the highest and the FTD-caregivers estimated a significantly higher burden in the FTD group compared to the ALS and IPS groups; FTD patients presented with a significantly lower level of disease insight, compared to the ALS and IPS groups. In all three groups, family and social support influenced sTBS.

**Discussion:** Both ALS and IPS groups had a good disease insight and a subjective burden comparable to that of one another, whereas the FTD group, displayed the highest level of self-assessed disease burden but also the lowest level of disease insight in this study. Cognitive and behavioural impairments and depressive symptoms negatively influenced sTBS in ALS and IPS groups. Only in the IPS group, physical health and disease duration played a critical role in determining their subjective burden, whereas in ALS and FTD, this was not the case.

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**COG-13 A role of psychological distress/trauma in ALS etiopathology?**

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**Background:** Recent research highlighted the crucial role of psychological distress and traumatic experiences in neurological diseases (1). This idea is consistent with psychosomatic transition into which framework Alexander (2) delineated two pathways of peculiar neurophysiological dysfunctions, indicating how the nature of traumas impacts different impairments/illnesses both at central and peripheral nervous system level. Mainly in terms of traumas involving the fight-or-flight system that impact the sense of agency/helplessness.

**Introduction:** ALS’ morphofunctional picture reveals interesting analogies with cerebral alterations caused by traumatic life events, such as excitotoxicity (3) and functional alterations of the right hemisphere (4). We hypothesized that in the time span in which ALS silently onsets, i.e. 5–15 years before manifest onset (5), patients might have experienced more traumas characterized by lower agency, lower elaboration and higher intensity than controls. Such vulnerability factor might add to the existing ones for ALS (e.g., cigarette smoking and physical trauma) (6).

**Methods:** We retrospectively assessed traumatic events with the TEC (7) in 71 patients and 76 controls. Patients with neuropsychological impairments or psychopathological manifestation were excluded from the sampling. A subsample of 31 patients and 31 controls accepted to be interviewed for a qualitative analysis of their traumas.

**Results:** Mixed-effects regression model analyses on the TEC showed that patients had significant more traumas than HC, \( \beta = 0.05 \pm 0.02 \) (\( p < 0.01 \)), during lifespan. Moreover, patients experienced traumas with a higher intensity, \( \beta = 0.13 \pm 0.06 \) (\( p = 0.02 \)). Post-hoc Tukey test on the subsequent interviews revealed that patients experienced a significant lower agency (\( p = 0.01 \)) than HC in the temporal window of 5–15 years before the diagnosis (patients’ M = 1.88, SD = 0.90; controls’ M = 2.7, SD = 0.65).

**Discussion:** Although the limitations imposed by the nature of retrospective studies, we hoped that our findings contribute to delineate one of the factors predisposing to ALS. Such an etiopathological deepening, if confirmed by further studies, might have a twofold value, both from a research perspective and at a clinical level for the prevention of the disease, hence suggesting important insights for clinical interventions with ALS patients.

**COG-14 Clinical features at onset and longitudinal trajectories of decline in MND patients with cognitive-behavioral impairment**

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**Background:** Cognitive and/or behavioral disturbances are frequently observed in motor neuron diseases (MNDs). However, it is still unclear whether MND patients with cognitive and/or behavioral impairment (MND-CBI) exhibit unique clinical profiles compared to MND patients with a pure motor syndrome (MND-motor).

**Objective:** To explore clinical differences between MND-CBI and MND-motor patients both at disease onset and over disease course.

**Methods:** 61 MND patients were followed longitudinally with mood and behavioral evaluations approximately every 3
months, and with cognitive and motor examinations approximately every 6 months, for up to 15 months. Cognitive and behavioral alterations were assessed using the Edinburgh Cognitive and Behavioural ALS Screen (ECAS), raw scores were age and education corrected using corresponding normative values. Mood disturbances were evaluated using the Hospital Anxiety and Depression Scale (HADS) and motor impairment was evaluated using the ALS Functional Rating Scale Revised (ALSFRS-r) and its rate of progression at baseline. Individual slopes of decline for each clinical measure were generated and Chi-squared and Mann-Whitney U tests were then used to compare baseline and longitudinal features between MND-CBI and MND-motor patients.

**Results:** According to current criteria, 32.79% of patients (N = 20) were classified as MND-CBI, while the remaining 67.21% of cases (N = 41) as MND-motor. At baseline, no significant differences were observed between the two groups in terms of demographic and mood features, while MND-CBI cases showed higher baseline ALSFRS-r progression rate. MND-CBI patients performed poorer than MND-motor cases also in ALS non-specific measures (ALS non-specific total score p = 0.04, memory p = 0.05 and visuospatial abilities p = 0.01). 59.01% of patients (N = 36) had at least one follow-up at the time of the investigation. Longitudinally, no significant differences were observed in the rates of motor decline. 32.14% of MND-motor patients converted to MND-CBI and 25% of MND-CBI cases developed additional behavioral symptoms. Both groups declined in ALS specific functions, however, a more severe worsening was selectively observed in MND-CBI (p = 0.02), mainly driven by a significant deterioration in the executive domain (p = 0.003). Both groups declined in ALS non-specific functions, MND-motor patients in the memory domain and MND-CBI patients in the visuospatial domain, but these differences did not reach significance. Mood disturbances ameliorated in MND-CBI and worsened in MND-motor cases, without a significant effect.

**Discussion:** This study provides novel insights into the clinical profile of MND-CBI. Our results suggest partially distinct onset features and longitudinal trajectories of extra-motor disturbances in this MND phenotype compared to pure motor cases.

**COG-15 The incidence of depression and quality of life in caregivers of patients with amyotrophic lateral sclerosis in Germany and Poland**

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**Background:** Foregoing observations show that ALS is a family disease affecting everyone in the patient’s environment. Improving the wellbeing of the patient’s caregivers has a positive impact on the patient’s quality of life. Identifying modifiable factors affecting the caregivers’ wellbeing may improve the overall care in ALS (1).

**Objectives:** To compare the quality of life and frequency of depression in caregivers of ALS patients in two neighboring European countries, Poland and Germany (n = 164), and to analyze correlation with demographic and/or clinical factors related to both caregivers and patients.

**Methods:** To determine the quality of life we used Anamnestic Comparative Self-Assessment (ACSA) (2) and Quality of Life in Life-Threatening Illness - Family Carer Version (QOLLTI-F) (3), and for depression the ALS Depression Inventory - 12 items (ADI-12) (4). Patients’ clinical stage was assessed with ALS Functional Rating Scale - revised (ALSFRS-R) (5), Edinburgh Cognitive and Behavioral ALS Screen - Behavioral Score (ECAS-BS) (6), disease duration and time from diagnosis.

**Results:** Caregivers from both countries reported a comparable and moderately good quality of life (p = 0.16 – QOLLTI-F, p = 0.22 – ASCA). A better caregiver’s QOLLTI-F was associated with a higher ALSFRS-R (p < 0.003) and a lower cognitive impairment (p < 0.005) of the respective patient. Age of caregivers negatively correlated with ASCA (p < 0.045), especially in the German group (p = 0.009). Depression was significantly more frequent among Polish compared to German caregivers (chi² = 4.899, p < 0.027). In both countries, the severity of depression in the caregivers was associated with a worse functional state of the patient (p < 0.004 vs. p < 0.05, respectively).

**Discussion:** Despite the extremely unfavorable prognosis and difficulties faced by the caregivers, most of them maintain a moderately good quality of life. We did not show significant differences between countries, despite different cultural background or financial and social assistance systems. In contrast, the depression rate among the caregivers was significantly higher in the Polish sample reflecting country-specific rather than disease-specific conditions.

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**Acknowledgements**

The authors thank the patients and caregivers from Germany and Poland for participating in this study. This is part of an EU Joint Programme – Neurodegenerative Disease Research (JPND; 01ED1405) project.
COG-16 Exploring the acceptability of CALM, an online self-help psychological intervention for people with MND and family members

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Background: There is an urgent need to develop and test psychological interventions to support people with MND and family members. We developed an online self-help psychological intervention called Coping and Living well with MND (CALM) to help people with MND and family members deal with the emotional impact of MND. This intervention/website draws on techniques from cognitive behaviour therapy (CBT), acceptance and commitment therapy (ACT) and mindfulness and was developed using insights from qualitative research with people with MND and family members.

Methods: The website was evaluated in two phases. In Phase 1, we conducted ‘think-aloud’ interviews with people with MND (n=9) and family members (n=9) to get detailed feedback on the acceptability of the content and presentation of the website. Findings from Phase 1 were used to iteratively refine the website. In Phase 2, different people with MND (n=18) and family members (n=9) were given access to the website and asked to use it and try out the suggested activities. After 6 weeks of using the website, we conducted interviews to explore their experiences. Interview data was analysed using thematic analysis.

Results: In Phase 1, feedback included optimising aspects of the language and presentation of the website and implementing minor changes to the activities and activity instructions. These changes were necessary to improve inclusivity for people with varying levels of physical and cognitive ability whilst ensuring it was also suitable for family members. The changes also helped reduce the possibility of the content triggering frustration and sadness or seeming off-putting or unfeasible. In Phase 2, the website was described as accessible and easy to use. Mindfulness activities, strategies to deal with difficult thoughts, and suggestions to do activities to stay positive were reported as useful to deal with difficult emotions. Some of the tips and activities were difficult for people with more severe symptoms and people whose symptoms were progressing quickly. There was mixed feedback about the use of images and videos, where some wanted more images or videos of people with MND and family members sharing their experiences, but some felt images or examples of people with certain symptoms or using certain equipment could be too confronting.

Discussion: An online intervention for emotional support can be useful for people with MND and family members. There is some variability in who finds online psychological support useful, and at what stage in the disease this support is useful. Further research is needed to determine how effective this intervention is for reducing distress and improving quality of life, and how to tailor online psychological support for people with MND and family members.

Acknowledgements
This work was funded by the Motor Neurone Disease Association, UK.

COG-17 ALS Focus caregiver needs survey results: what matters most to ALS caregivers

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Background: ALS Focus is a patient- and caregiver-led survey program designed to measure the needs, preferences, and experiences of people with ALS and their caregivers across the disease. Given the critical role that caregivers play in the United States, ALS Focus set out to measure—across a large sample of current and past ALS caregivers—which programs and tools are critical to ALS caregivers, their top concerns, and quality of life.

Methods: The ALS Focus Caregiver Needs survey took place from January to April 2021. Current and past caregivers of people with ALS opted into this self-reported and de-identified online survey through the ALS Focus survey platform (www.alsfocus.org). Survey measures asked caregivers about time spent providing care, use of ALS programs and trainings, the top five tools needed in their caregiving roles, and their top five concerns. Current caregivers also reported how prepared they felt for future caregiving responsibilities and rated their quality of life overall. The ALS Focus Patient and Caregiver Advisory Committee (PCAC), industry experts, and Care Services leaders from The ALS Association consulted on survey development. ALS Focus data collection methods were presented previously (1) and are described at www.als.org/ALS-Focus.

Results: In total, 324 current caregivers and 287 past caregivers participated in the Caregiver Needs survey. Results showed ALS caregivers dedicate a lot of time to providing care, with 68% saying they spend over 30 hours per week. Although few participants were professional caregivers (6%), for most, the time spent caregiving was equivalent to a part-time or full-time career (76%). Home visits from nurses and occupational or physical therapists ranked as the most highly used (53%) and highly needed (49%) ALS program. Fewer caregivers reported utilizing trainings on general caregiving (34%), but these trainings ranked second (42%) in terms of mattering most to caregivers in their roles. Most often, caregivers said their family and loved ones’ wellbeing was a top concern (61%). Time for self-care, decreased engagement in enjoyable activities, and depression also rankly highly among caregivers’ concerns. Among current caregivers, 46% reported feeling unprepared for changes in caregiver responsibilities as ALS progresses, and nearly 37% rated their quality of life on the lower half—or poor—of a 10-point scale. We discuss
implications for how to serve ALS caregivers and ensure their wellbeing as they focus on the wellbeing of people with ALS.

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COG-18 Subjective health perception prioritizes psychological well-being over physical function as ALS advances

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Background: Despite declining physical function, patients with ALS report relative preservation of overall individual quality of life (QoL). This paradoxical finding is attributed to psychological adaptation to deficits with reprioritization of the individual construct of QoL. A similar adaptation could also affect subjective health perception (HRQoL).

Objective: The aim of this cross-sectional study was to examine reprioritization of factors that determine HRQoL with disease progression using structural equation modeling (SEM).

Methods: Consecutive ALS patients seen in the course of routine care self-reported ALSFRS-R (a measure of bulbar, motor, and respiratory function), PHQ-9 (a measure of depression), and EQ-5D-3L (a 5-dimensional utility index that also includes a visual analog scale [EVAS] asking about health perception on a line segment). ALS was staged by the FT9 method and classified into early (FT9 stages 0, 1 and 2) and late (FT9 stages 3 and 4) disease. Measurement models were constructed for the latent factors of physical and psychological well-being (PHY and PSY respectively), with appropriate indicators chosen from self-reported measures. Model fit was assessed by standard metrics. EVAS was used as the overall measure of HRQoL. Weights of PHY and PSY on EVAS for early and late disease were estimated to examine reprioritization.

Results: Of the cohort of 578 patients (mean age 61.5 ± 11.9, 59% male), 423 (73%) were early and 155 (27%) were late disease. Mean ALSFRS-R, PHQ-9, EQ-5D index utility and EVAS for early vs late stage were 39.9 ± 4.7 vs 27.1 ± 7.2, 5.9 ± 5.2 vs 10.2 ± 5.9, 0.72 ± 0.18 vs 0.51 ± 0.21, and 63.5 ± 20.6 vs 48.6 ± 20.9, respectively ($p < 0.001$ for all). A measurement model was established with good model fit (RMSEA = 0.054, CFI = 0.958, TLI = 0.941, SRMR = 0.038), where latent variables were defined as PHY (ALSFRS-R and EQ-5D questions 1–3) and PSY (PHQ-9 items and EQ-5D question 5). A structural model was estimated with PHY, PSY, and pain (marked by question 4 of EQ-5D) predicting HRQoL. In unadjusted models, standardized weights of PHY and PSY on HRQoL in early disease were 0.42 (standard error = 0.08) and 0.19 (0.07) respectively, whereas for late disease they were 0.28 (0.09) and 0.41 (0.08). Importantly, PHY and PSY were significantly correlated in early but not in late disease.

Discussion: This study confirms the hypothesis that health perception (HRQoL) is more representative of psychological well-being and less representative of physical function as disease advances, in parallel with overall QoL. The effect of disease duration on this adaptation is explored. Subjective health status may differ from health utility, and may be of importance for health technology assessment. Greater allocation for psychological health would be the most effective strategy to maximize subjective health status as ALS advances.

COG-19 Schizotypal traits across the frontotemporal dementia–motor neuron disease spectrum: pathomechanistic insights

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Background: Psychiatric symptoms are highly prevalent across the frontotemporal dementia-motor neuron disease (FTD-MND) spectrum, present in up to 50% of cases (1). Despite this, schizotypal personality traits that are characterised by subclinical psychotic-like perception, thoughts and behaviours and have been independently associated with an increased risk for the development of clinical psychosis, poor psychosocial outcomes and functional impairment, have never been examined (2).

Objective: The current study aimed to address this gap in knowledge by providing the first, detailed examination of all subtypes of schizotypal traits across the FTD-MND spectrum, as well as exploring the neural underpinnings to characterize potential underlying mechanisms.

Methods: 109 participants were recruited to the study (37 MND, 10 FTD-MND, 45 behaviour variant FTD, and 17 age-, sex- and education-matched healthy controls) and were administered the comprehensive 74-item Schizotypal Personality Questionnaire to assess Schizotypal traits (3). The association between changes in grey matter volume density and severity of schizotypal traits was examined using voxel-based morphometry (VBM) analysis on T1-weighted structural magnetic resonance imaging.

Results: Relative to controls, pervasive schizotypal personality traits across the domains of positive and negative schizotypy and disorganised thought disorders were present across all patient groups (all $p < 0.001$), confirming a wide spectrum of subclinical schizotypal symptoms, beyond classic psychotic symptoms, across the FTD-MND continuum. Atrophy in frontal, anterior cingulate and insular cortices, and caudate
and thalamus was implicated in positive schizotypy, while integrity of the cerebellum was associated with disorganised thought disorder traits (corrected for cluster-extent multiple comparisons at \( p < 0.05 \), with a cluster-forming threshold of \( p < 0.001 \)).

**Discussion:** The frontal-striatal-limbic regions underpinning manifestation of schizotypy in the FTD-MND spectrum are similar to those widely established in schizophrenia neuroimaging research. This finding expands the concept of a psychiatric overlap in FTD-MND and schizophrenia-related conditions and suggests potentially common underlying mechanisms involving disruptions to frontal-striatal-limbic networks in the emergence of psychotic symptoms. This also holds implications for future psychiatric and neurodegenerative disorder research, warranting a transdiagnostic approach for investigation and interpretation of the clinical phenomena related to psychosis.

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**Acknowledgements**

This work was supported by funding to Forefront, a collaborative research group dedicated to the study of frontotemporal dementia and motor neuron disease, from the National Health and Medical Research Council of Australia program grant (#1037746) and the Australian Research Council Centre of Excellence in Cognition and its Disorders Memory Program (#CE110001021).

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**COG-20 Efficacy of a mindfulness intervention for people with motor neurone disease and their family caregivers: comparison of those with high and low depression**

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**Background:** People with MND and their family members (FmMND) face significant psychological adjustment challenges related to increasing disability and a markedly shortened lifespan, including increased depression and anxiety. It is unclear whether psychological interventions should be targeted to those showing most distress, as is typical in resource stretched services, or offered more broadly, including to those with lower distress levels. Given the challenge of MND, even those with no overt mental health symptoms may benefit from a psychological intervention to support emotional wellbeing and psychological resilience. Mindfulness has been shown to be efficacious across clinical and non-clinical populations, and has demonstrated efficacy for those with MND (1). In MND, it is not known whether this intervention may differentially affect those with mental health symptoms or without. **Objectives:** The aim of this research was to identify people with MND and FmMND with high versus low depressive symptoms, and to compare these groups in relation to (i) baseline demographic and psychological features and (ii) engagement with and response to a mindfulness intervention.

**Methods:** Potential participants with early to middle stage MND were identified from the patient database at a multidisciplinary MND clinic. Fifty seven eligible patients and FmMND agreed to participate in a fortnightly four-session mindfulness program adapted for MND. A single arm waitlist design was employed, with assessments pre-waitlist (T1), pre-intervention (T2) and post intervention (T3). Measures examined depression and anxiety, demoralisation, quality of life, mindfulness skills, and benefit-finding. Assessments of between-session practice were also included. Analyses involved parametric and non-parametric statistics.

**Results:** Forty participants (22 male, age: \( M = 63 \text{yrs} \), range 20–88) completed the program (retention rate =70%). Based on the modified Hospital Anxiety and Depression Scale scores (2), participants were divided into high depression \((n = 11)\) and low depression \((n = 28)\) groups. Group comparisons showed no differences in demographic features or disease status and strong differences in baseline psychological characteristics. The groups showed different patterns of engagement of in mindfulness practice over the course of the intervention. In response to the intervention, the high depression group showed decreased anxiety \((p < 0.05)\) and positive changes in benefit-finding and quality of life \((p < 0.1)\), while change was not evident in the low depression group.

**Discussion:** Measurable benefit from a mindfulness-based intervention was shown amongst those with increased depression. Where there are resource constraints in the provision of psychological interventions, they should be targeted towards those with mental health symptoms. A longer follow-up period would be needed to examine whether mindfulness interventions are protective of future distress in later stages of MND.

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**COG-21 Clinical relevance of dysgraphic features in western non-aphasic ALS patients**

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1. COG-21 Efficacy of a mindfulness intervention for people with motor neurone disease and their family caregivers: comparison of those with high and low depression

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Background: Up to 35–40% of non-demented ALS patients show oral language impairment within the spectrum of frontotemporal degeneration (1), whose diagnostic and prognostic entailments have been highlighted (2). Central dysgraphic features, reflecting actual language deficits, have been also described (3), although their clinical relevance in ALS is yet to be proved, especially for western languages, as current literature is mostly focused on Japanese patients (2,3).

Objectives: To describe clinical and cognitive features of clinically non-aphasic ALS patients displaying writing errors (WE).

Methods: Eight consecutive ALS patients with no other major co-morbidities underwent clinical assessment (ALSFRS-R; MiToS and King’s staging) and both brief and multi-domain cognitive screening: the ALS Cognitive Behavioural Screen (ALS-CBS) (4) and the Edinburgh Cognitive and Behavioural ALS Screen (ECAS) (5). Writing was assessed via a writing-to-dictation task of an ad-hoc list of words and non-words (29 stimuli).

Results: Mean age of the 8 patients was 61.3 ± 12.5; mean education 13.5 ± 6.1; four were males. Mean disease duration was 39.4 ± 27.4 months. ALS phenotype was heterogeneous (3 classical; 2 flail leg; 1 pyramidal; 1 PLS; 1 respiratory); six patients had a spinal onset; bulbar signs at evaluation were present in four. Mean ALSFRS-R was 32.5 ± 5.4. Median MiToS (3 classical; 2 flail leg; 1 pyramidal; 1 PLS; 1 respiratory); six patients had a spinal onset; bulbar signs at evaluation were present in four. Mean ALSFRS-R was 32.5 ± 5.4. Median MiToS and King’s stages were 1 and 3, respectively. WE were present in 6/8 patients (Mdn = 3, IQR = 2; range: 0–7) that could not be accounted for motor disabilities. No defective ALS-CBS scores were detected. Patients were subdivided according to the number of WEs: WE+ (making ≥ 3 errors), and WE- (making < 3 errors). Among WE+ (N = 4), three had below-cut-off ECAS-total and -ALS-Specific scores. Among WE- no patient had impaired ECAS-total/-ALS-Specific scores. Bulbar sign prevalence was 3/4 in WE+ vs. 1/4 in WE-patients. King’s stage 3 was more represented in WE+ patient (3/4 vs. 1/4 WE-patient in King’s stage 4). Phenotype, onset and MiToS across-group distribution revealed no specific frequency patterns.

Discussion: Some western non-aphasic ALS patients show central dysgraphic features, in agreement with the literature regarding eastern patients (3). These patients present with executive/language dysfunctions that are detectable by multi-domain screens (ECAS). Therefore, writing-to-dictation tasks might be valid to test for frontotemporal involvement in this population, particularly in severely dysarthric patients. WE in ALS patients are associated with bulbar involvement and advanced disease (as assessed by King’s staging).

Acknowledgements

The Authors thanks Prof. Claudio Giuseppe Luzzatti and Prof. Eleonora Caticala for providing us with test materials adopted in this work.

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COG-23 Perceived social isolation is associated with neurobehavioral functioning in patients with ALS

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Background: Perceived social isolation, also termed loneliness, impacts on neurobiological architecture, ensuing significant consequences on mental and physical health. In elderly people, it represents a risk factor for dementia. In ALS, loneliness has been found to be related to poor psychological health, however, the possible backlash of loneliness on behavioural and cognitive profiles of ALS has been poorly investigated.

Objectives: To explore the association of loneliness with behavioural and cognitive symptoms of frontotemporal syndromes of ALS.

Methods: Loneliness was measured using the 3-item UCLA Loneliness Scale (range: 0–6 points; no loneliness: 0 points, mild/moderate loneliness: 1–3 points, high loneliness: 4–6 points) in 206 consecutive ALS patients. The Edinburgh Cognitive and Behavioural ALS Screen, the Story-based Empathy Task and the Ekman 60-Faces Test were used to evaluate cognitive efficiency and social cognition. Behavioural symptoms were measured with the Frontal Behavioural Inventory (FBI-ALS) and the Dimensional Apathy Scale (DAS). Self-reports of mood (Hospital Anxiety and Depression Scale for motor neuron disease - HADS-MND), alexithymia, Difficulty in Emotional Regulation (DERSF) and quality of life (QoL) were also used. Spearman rho and Jonckheere-Terpstra test examined factors mainly associated with UCLA scores. Significance level was set at <0.001.

Results: In our cohort, 126 patients (62%) reported no loneliness, 70 (33%) were classified as low/moderately lonely; and 63 (31%) were classified as highly lonely. UCLA scores were significantly associated with FBI (Z0.413), but not with cognitive abilities including DAS (Z0.402), anxiety (Z0.447) and QoL scores (Z0.338), DERS (Z0.457), anxiety (Z3.855), alexithymia (Z3.852), DERS (Z5.793) and QoL (Z5.038).

Discussion: Our findings indicated that, in ALS, the satisfaction of social environment is associated with a sense of life well-being that is not limited to the functional motor status. Loneliness was strongly related to neurobehavioral functioning and not with cognitive abilities. Depressive mood, anxiety, disinhibition and apathy were higher in lonely patients. Feelings of loneliness were also accompanied by subjective emotion dysregulation and poor emotional awareness, but not with empathy and emotion recognition dysfunctions. The relationship of loneliness to the behavioural and psychiatric symptoms is likely bidirectional; in some situations, it may act as a risk factor and in others it may be the consequence of these symptoms. Paying attention to social isolation in patients with ALS will help clinicians to intervene at an early stage.

Acknowledgements
This work has been supported by Fondazione Regionale per la Ricerca Biomedica, Regione Lombardia (TRANS-ALS: 2015-0023) and by Fondo Europeo di Sviluppo Regionale, Regione Lombardia (POR FESR 2014-2020; 1157625).

COG-24 Current practices of using mental capacity assessments in the clinical care of MND patients in the UK

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Background: The issue of mental capacity is particularly salient in people with MND (pwMND) given that up to 50% experience changes in cognition and/or behaviour (1). The life-limiting nature of MND means that important and difficult decisions are required to be made throughout the course of the disease (2). Healthcare professionals (HCPs) must therefore judge patients’ capacity to make specific decisions. However, it is unknown how capacity assessments are used in the clinical care of pwMND in the UK.

Objectives: To investigate HCPs current practices of using mental capacity assessments in the clinical care of pwMND in the UK.

Methods: HCPs working in UK MND clinics were invited via email to participate in an online survey about their current practices of using capacity assessments in pwMND. Items specifically explored the characteristics of the context and frequency of capacity assessments and features of the assessment process.

Results: 97/160 responded to the survey from England (73.9%), Scotland (14.1%), and Wales (12%). 49/86 (57%) were involved in capacity assessments of pwMND. Of these, around 90% had encountered pwMND who lacked capacity to make specific decisions. Such decisions included those relating to: gastrostomy, non-invasive ventilation, accommodation, personal care, and legal issues (e.g. power of attorney). Assessments typically took place in the patient’s home or within a hospital out-patient setting, and the time taken to administer assessments varied. The majority of respondents completed less than 10 capacity assessments per year.
respondents checked the following optional responses: general discussion with the patient/carer (29%), clinical judgement based on a semi-structured interview about the specific decision (28.0%), using a standardised assessment/tool (14%), and clinical judgement based on discussion within clinic (13%). If a cognitive screening indicated cognitive impairment, only 55.6% said that this would prompt a capacity assessment to be carried out if a specific decision needed to be made. When questioned if life-prolonging interventions would still be offered to those with cognitive impairment, around a third said yes, whilst around two-thirds said that it would depend (most often on the nature and degree of cognitive impairment).

**Conclusions:** The study provides insight into the current practices of using capacity assessments in the clinical care of pwMND in the UK.

**Acknowledgements**

This study was funded by the SPRINT-MND/MS PhD studentship.

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**COG-25**

Prominent upper motor neuron dysfunction is associated with the presence of behavioural impairment in patients with amyotrophic lateral sclerosis

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**Background and aim:** Increasing evidence shows that approximately half of patients with amyotrophic lateral sclerosis (ALS) displays cognitive (ALS-cognitive) or behavioural (ALS-behavioral) impairment, or both (ALS-cognitive and behavioral). (1) Aim of our study is to assess whether the burden of upper (UMN) and lower motor neuron (LMN) involvement is associated with the presence of cognitive and behavioural impairment.

**Methods:** A single-centre retrospective cohort of 110 Italian ALS patients has been evaluated to assess correlations between motor and cognitive/behavioral phenotypes. UMN regional involvement was measured with the Penn Upper Motor Neuron Score (PUMNS), (2) while LMN signs were assessed using the Lower Motor Neuron Score (LMNS) (3). The Edinburgh Cognitive and Behavioural ALS Screen (ECAS) (4) - Italian version and the Frontal Behaviour Inventory (FBI) were administered to evaluate patients’ cognitive and behavioural profile.

**Results:** PUMNS at first visit was significantly higher in behaviourally impaired ALS patients (ALS-bi and ALS-cbi) compared to behaviourally unimpaired individuals (ALS and ALS-cs) (9.90 vs 6.97, p = 0.014). Concerning the different FBI subdomains, higher PUMNS correlated with the presence of Apathy, Emotive Indifference, Inflexibility, Inattention, Perseveration, and Aggressiveness.

**Conclusion:** To our knowledge, this is the first study showing that a clinical prominent UMN dysfunction is associated with a more significant behavioural impairment in ALS patients, suggesting the hypothesis of a preferential spreading of the pathology from the motor cortex to the ventromedial prefrontal and orbitofrontal cortex in this group of patients.

**COG-26**

Towards family-centered ALS care: development of a guide for ALS care professionals on how to support parents and children in families living with ALS

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**Background:** ALS affects not only patients but also the families surrounding them, especially when dependent children are involved. Efforts are needed to better support children in families living with ALS, both directly and through strengthening parents in their parental role. ALS care teams may play an important role in empowering parents and children in
families with ALS. However, due to lacking guidelines, ALS professionals are often uncertain about how they can provide optimal care to parents and children in families living with ALS.

**Objectives:** To provide ALS care professionals with guidance on how to support parents and children in families living with ALS.

**Methods:** A practical guide for family-centered ALS care was developed, based on (1) a systematic review of the literature on support needs of families with ALS and other life-limiting illnesses; (2) experiences, struggles and needs of parents and children living with ALS, explored through semi-structured interviews with 21 parents (8 with ALS) and 15 children; (3) a survey among ALS care professionals (N = 65) to map the support ALS care professionals provide to families living with ALS and to identify their needs, struggles and knowledge gaps with regard to ALS care for families; and (4) input from a workgroup including ALS care professionals, ALS patients and relatives.

**Results:** A practical guide incorporating generic and specific guidance was developed to facilitate ALS care professionals in supporting parents and children in families with ALS. Generic guidance involves education and collaboration between family and professional. Specific guidance is clustered around four themes: (1) awareness of the family as a system, (2) attention to the parental role, (3) psycho-education of parents, and (4) involvement of children in ALS care. The guide contains practical recommendations and tools for identifying family support needs, starting a dialogue about parenting issues, assessment of family functioning, and referral.

**Discussion:** The findings have important implications for current ALS care practices. The newly developed guide for family-centered ALS care may contribute towards improving the quality of care provided to families living with ALS. Although this guide has been developed in the Netherlands, the themes identified may be applicable in other countries/cultures as well.

**Acknowledgements**

We thank the ALS Foundation Netherlands for financially supporting this study. Furthermore, we thank the ALS care professionals, patients and relatives who participated in the interviews and/or workgroup.