COG-01: A different cognitive and behavioral profile in ALS patients with or without C9orf72 expansion

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Objectives:
The aim of this study was to analyze whether ALS patients with C9orf72 expansion showed a different profile of cognitive and behavioral domains compared to patients without C9orf72 expansion (a) at the same level of motor impairment, classified according to King's staging system, and (b) at the same degree of cognitive and behavioral deficit, classified according to the revised ALS-FTD Consensus Criteria.

Material:
We considered 741 ALS patients, consecutively seen at the Turin ALS center in the period 2010-2018, who underwent both cognitive/behavioral and genetic testing. Patients were diagnosed according to El Escorial revised criteria. Kings' staging and genetic analysis at time of cognitive testing were collected for all patients. ALS patients underwent a neuropsychological battery selected according to the ALS-FTD Consensus Criteria.

Methods:
Comparisons were performed on age-, sex-, and education-corrected scores. The Mann-Whitney U test was used for comparisons. First, comparisons were performed for each King's stage, independently from their level of cognitive status. Second, we matched the results of cognitive tests for each behavioral and cognitive level, merging the intermediate cognitive categories. Third, we confronted the number of ALSC9+ vs. ALSC9- cases who showed an impairment in each neuropsychological domain.

Results:
ALSC9+ patients were younger than ALSC9- at all levels of cognitive impairment. ALSC9+ patients had significantly lower scores in tests exploring executive functions and verbal memory both when classified as cognitively normal and when diagnosed in the intermediate cognitive categories. Considering the clinical perspective, ALSC9+ patients showed significantly lower scores compared to ALSC9- patients at King's stage 1 and 3 in almost all the examined neuropsychological domains, while at King's stage 2 ALSC9+ patients were more severely affected only in the verbal memory domain. Behavioral function was comparably impaired in the two cohorts.

Discussion:
Our data suggest that ALSC9+ patients show a comparatively different neuropsychological pattern. In general, they are more impaired in cognitive functioning than ALSC9- patients, especially in the domains of executive functions, visual memory, and verbal memory. Verbal memory emerged as a particularly vulnerable function in ALSC9+ patients, with lower scores even when they are still classified as cognitively normal. This could imply in ALSC9+ patients a 'cognitive' presymptomatic/subclinical condition characterized by lower performances at specific cognitive tasks when motor symptoms are already present.

Conclusion:
According to our data, it is conceivable that in some ALSC9+ patients a poor cognitive performance is already present in the early motor stages of the disease. Longitudinal studies are necessary to clarify whether this subclinical cognitive impairment in ALSC9+ patients will evolve to clinically overt dementia over time.

COG-02: ALS Parents & Kids Support: Lived experiences and support needs of parents and children in families with ALS

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Background:
Amyotrophic lateral sclerosis (ALS) affects not only the patient but also the family that surrounds him or her. Families with children deserve special attention. When a parent is diagnosed with ALS, attention often turns to how the children in the home are coping. Both the parent living with ALS and the well-parent worry about the impact of ALS on their children's lives and are uncertain how to optimally support their children. Despite previous research highlighting the vulnerability of children of a parent with ALS in terms of mental health, behavioural problems and quality of life, children are, as yet, often overlooked by caregivers when it comes to providing information and support. Efforts are needed to protect children throughout the illness trajectory and to empower parents to support their children in dealing with the diagnosis, the physical deterioration of the parent and the prospect of parental loss. To this end, it is crucial to have a better understanding of parents' and children's specific support needs. The current literature base focuses primarily on care needs, ignoring parent's and children's needs in the context of ALS.

Objectives:
To explore the lived experiences and support needs of parents and children in families living with ALS.

Methods:
Semi-structured interviews were conducted, either via telephone or online, with 6 parents with ALS, 11 well parents and 15 children/adolescents with a parent with ALS (age range: 13-23 years). Interview data were analyzed using qualitative content analysis.

Results:
With regard to parent's and children's lived experiences, two major themes were identified: (1) lived experiences of families with ALS, including distress towards self vs. others, changes in family dynamics and relationships, changes in the home environment, and family-level vs community-level involvement of children in ALS, and more specifically (2) experiences of parenting with ALS, including parental sense of competence, parental identity, role changes and parenting strategies. When addressing the support needs of parents and children in families with ALS, two additional main themes emerged concerning: (1) types of support needed, including emotional, practical, informational and psychological support, and (2) resources for support, including personal, social, informational and healthcare resources.

Discussion and conclusions:
This study not only corroborates previous research but also adds to the empirical knowledge base on parental and children's support needs in the field of ALS. The findings have important implications for current practices in ALS care. The themes identified in this study may guide the development of tailored support services for ALS families with children.

COG-03: Amyotrophic Lateral Sclerosis Patients' Socioecological Resilience, Self-Determination, and Decision-Making for Life-Sustaining Treatments

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Background:
Sixteen percent of people with ALS (pALS) get tracheostomies, and only 38% are planned (Ceriana et al., 2017), suggesting that many pALS' decisions for life-sustaining treatments are not fully self-deterministic. While researchers have examined resilience and self-determination in mental health and chronic illness, none have researched these variables with pALS from a socioecological framework.

Objectives:
This cross-sectional correlational mixed-methods study explored and examined the relationships between pALS' socioecological resilience, self-determination, and decision-making for life-sustaining treatments.

Methods:
Soliciting study participants randomly through the National ALS Registry, 197 pALS provided informed consent and completed online surveys. The participants answered 17-items from the Adult Resilience Measure-Revised, 21-items from the Basic Psychological Need Satisfaction Scale, four yes/no questions about their decision-making for life-sustaining treatments, 18-demographic items, and eight qualitative items surrounding resilience and self-determination. Thematic and content analyses were performed with the qualitative data. Binary logistic regression tests were done to determine the relationships between the primary quantitative variables. Bivariate correlation tests were also performed to determine the relationship between the independent variables.

Results:Qualitative content and thematic analysis revealed that pALS' perceived burdens, disease progression, functional abilities, profound loss, quality of life, adaptability, resources, relationships, and environmental and supernatural forces contributed to their decision-making for life-sustaining treatments. Seven transcendent themes were observed across the 12-qualitative items. Binary logistic regressions showed no significant relationships between socioecological resilience (ρ = .086, ρ = .232), self-determination (ρ = .177, ρ = .674), and decisions for tracheostomies and PEGs, respectively. Significant relationships were found between the covariates, including age (ρ < .001, ρ = .028, ρ = .026), gender (ρ < .001), military veteran status (ρ = .003, ρ = .002), and disease progression (ρ < .001) and decsions for tracheostomies and PEGs, respectively. Bivariate correlation test results showed a strong positive relationship between pALS' socioecological resilience and self-determination (r = .715, ρ < .01).

Discussion:
The joint display table manifested four points of convergence, which showed that age, military veteran status, disease progression, and tensions related to pALS' circumstances contributed to their decision-making for life-sustaining treatments. Positive social change implications include establishing an ecological decision-making model to improve social work services and empower pALS' decision-making. The findings also demonstrate empirical rationales for increased socioeconomic resources to support pALS' decisions for life-sustaining treatments.

References:
Ceriana, P., Surbone, S., Segagni, D., Schreiber, A., & Carlucci, A. (2017). Decision-making for tracheostomy in amyotrophic lateral sclerosis (ALS): A retrospective study. Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration, 18(7-8), 492-497. https://doi.org/10.1080/21678421.2017.1317812

COG-04: Associations between illness cognitions and health-related quality of life in the first year after diagnosis of amyotrophic lateral sclerosis

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Objective:
To describe illness cognitions among patients with amyotrophic lateral sclerosis (ALS), to study cross-sectional associations between illness cognitions and health-related quality of life (HRQoL) and to study the predictive value of illness cognitions measured shortly after the diagnosis for HRQoL at follow-up.

Methods:
Prospective longitudinal design. We administered Self-report questionnaires at study onset (n=72) and follow-up (n=48). Median follow-up period was 10.0 months. At baseline median ALS Functional Rating Scale-Revised was 43, median time since onset of symptoms was 13.6 months, 79% of patients presented with spinal onset. Illness cognitions Helplessness, Acceptance and Disease Benefits were measured with the Illness Cognitions Questionnaire (ICQ) and HRQoL with the ALS Assessment Questionnaire (ALSAQ-40). Correlational and regression analyses were used.

Results:
Patients experienced more Helplessness at follow-up. We found no significant changes in Acceptance or Disease Benefits at follow-up. In cross-sectional analyses, Helplessness was independently related to worse HRQoL at baseline (β= 0.44; p = 0.001) and Acceptance and Disease Benefits were independently related to worse HRQoL at follow-up (β = -0.17, p = 0.045) and β = -0.186, p = 0.03 respectively). Longitudinal analyses showed that, adjusted for disease severity at baseline, Helplessness at baseline was a predictor of worse HRQoL at follow-up (β=0.43; p=0.006). None of the illness cognitions were a significant predictor of HRQoL with adjustment for baseline HRQoL

Conclusion:
Helplessness was independently associated with HRQoL in the cross-sectional and longitudinal analyses. These results can help us identify patients shortly after diagnosis who might benefit from psychological interventions.

Kruitwagen-van Reenen ETH, Mwm P, van Groenestijn A, van den Berg LH, Jma VM. Associations between illness cognitions and health-related quality of life in the first year after diagnosis of amyotrophic lateral sclerosis. J Psychosom Res. 2020 May;132:109974. doi: 10.1016/j.jpsychores.2020.109974.

COG-05: Change over time and impact of apathy in amyotrophic lateral sclerosis

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Background:
Apathy can occur in up to 50% of people with amyotrophic lateral sclerosis (pwALS) and is associated with lower caregiver wellbeing and increased burden in ALS (1). Apathy is multidimensional and Initiation apathy (a lack of motivation for self-generation of thoughts and/or actions) is characteristic of ALS and may impact care (2,3). However, how different types of apathy change over the course of disease and how they affect the pwALS, and their carers has not yet been explored.

Objectives:
To determine how apathy subtypes change longitudinally, and their impact on patient quality of life, caregiver wellbeing and burden.

Methods:
50 pwALS and their caregivers were recruited from the CARE-MND platform (Scotland) and MND Care & Research Networks (England). Participants took part in interviews at 3 time points (baseline, 1st follow-up and 2nd follow-up) at 3-month intervals. They completed the Dimensional Apathy Scale (DAS), and assessments of depression, anxiety and emotional lability cognitive functioning and behavioural change (Edinburgh Cognitive & Behavioural ALS Screen) and functional disability (ALS Functional Rating Scale-Revised). ALS-specific quality of life (ALSSQoL), Zarit Burden Inventory (ZBI-SF), Carer Experience Scale (CES) and Carer Wellbeing (ICACAP-A) were also measured. Cross-sectional analysis and longitudinal analyses were performed on absolute change scores (difference between baseline and follow-up).

Results:
At baseline, Initiation apathy was most common (38%). Those with Initiation apathy had significantly lower scores on the ALSFRS-R (p<0.05), ICECAP-A (p<0.001) and ZBI-SF (p<0.01), than those with no apathy. For participants who completed all 3 visits (N=32), there were significant Initiation (p<0.01) apathy increase, with significant decrease in self-rated Emotional apathy (p<0.05) over time. There was also significant increase in ZBI-SF scores (p<0.001) and decrease in ICECAC-A (p<0.01) and CES (p<0.05) scores over time. There was no change in overall cognition, depression, anxiety and emotional lability. Increased absolute change scores in Initiation and Executive apathy were significantly correlated with increasing ZBI-SF scores (p's<0.05) and with decreasing overall ALSSQoL scores (p's<0.001). Additionally, decrease absolute change scores in Emotional apathy were significantly correlated with decreasing ZBI-SF scores (p<0.05).

Conclusions:
As disease progresses, there is an increase in behavioural impairment, specifically relating to Initiation apathy. Apathy subtypes (Executive and Initiation apathy) are associated with decrease in patient quality of life and caregiver burden. This suggests the importance of considering motivation of pwALS and caregivers wellbeing across the disease, with wider implications for practical management.

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COG-06: Cognitive changes in Georgian Amyotrophic lateral sclerosis Patients (ALS)

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Modern studies have shown strong link between amyotrophic lateral sclerosis and fronto-temporal dementia (FTD).

Purpose:
To study the cognitive changes in patients with ALS in Georgia.

Methods:
Totally 25 patients, aged 22 to 83, 13 male, 12 female were investigated with definite and probable ALS , defined by "El Escorial " criteria.   Cognitive changes defined with Addenbrooke's cognitive examination - ACE-III (attention , memory , fluency , language , visuospatial) and frontal behavioral inventory – FBI. Brain was visualized by MRI(1.5T), Electromyography (EMG) made in all patients. Probable ALS signs and Cognitive changes were researched in family history of all patients. Statistics performed by SPSS 20.

Results:
Frontotemporal lobar dementia was diagnosed in 5 (20%), 4 females,1 male, patients aged 55-67 years. These patients demonstrated cognitive impairment on ACE -III examination (mostly language, fluency) characteristic to Frontotemporal dementia and with radiological confirmation. All 5 patients had FTD spectrum disorders in family pedigree without history of ALS. Analysis of FBI showed loss of insight, indifference, and distractibility in 60% of FTD patients as well as perseveration, disinhibition and irresponsibility. Personal neglect and apathy were the most frequent negative symptoms shown in 40% of patients.

Multivariate logistic regression showed the significance of Autosomal dominant inheritance for developing FTD in Patients with ALS (p<0.01). Correlation was not found between the age of ALS patients and Cognitive decline (p<0.5).

Conclusion:
According to the present research in presumably sporadic ALS cases in Georgia there is a strong link for developing FTD. Further research is required.

Key words: dementia, motor neuron disease, inheritance, FTD

COG-07: The Amyotrophic Lateral Sclerosis Health Index (ALS-HI): A Measure of Prioritized Symptomatic Themes in ALS

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Background:
The ALS patient population experiences numerous clinical symptoms. In clinical trials, valid, relevant, and responsive patient-reported outcomes are needed to measure the diverse set of symptoms experienced by patients with ALS and support drug labeling claims (1).

Objectives:
1) To determine the prevalence and relative importance of ALS symptoms in a large population of patients.
2) To utilize this data in the development and validation of a sensitive, relevant, disease-specific patient-reported outcome measure for ALS, the Amyotrophic Lateral Sclerosis-Health Index (ALS-HI).

Methods:
We conducted a cross-sectional study of 497 participants with ALS enrolled in the Centers for Disease Control and Prevention National ALS Registry. Participants reported the relative importance and prevalence of 189 individual symptomatic questions representing different symptomatic themes. Responses were categorized by age, sex, education, employment status, respiratory status, and speech status. Questions were selected for inclusion in the ALS-HI based on their high importance, prevalence in ALS, and their potential to detect therapeutic change over time. In addition, we conducted cognitive interviews with ALS patients to determine the relevance, understanding, and usability of the ALS-HI and measured the consistency of each subscale included in the ALS-HI.

Results:
ALS participants provided over 89,000 responses to address the relative prevalence and importance of each ALS symptom. The highest prevalence symptomatic themes related to: inability to perform activities (93.8%), fatigue (92.6%), problems using hands and/or fingers (87.7%), limitations with mobility or walking (86.7%), and decreased performance in social situations (85.7%). Altogether, the ALS-HI consists of 15 subscales that measure a patient's overall disease burden while individually measuring a patient's perspective on specific aspects of their health. These subscales focus on the physical, emotional, social, and symptom-specific health of ALS. ALS-HI subscales were found to have a high interval consistency. During beta interviews; patients found the instrument to be highly relevant, user-friendly, comprehensive, and appropriate for measuring how they feel and function.

Discussion:
There are numerous symptoms that contribute to disease burden in ALS. ALS-HI is designed and validated to serially measure the symptomatic themes of greatest importance to ALS patients while providing a potential mechanism to record therapeutic benefit during future clinical trials.

References:
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COG-08: Cognitive performances in ALS patients depend on the lateralization of motor impairment

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Objectives:
In the present study we investigated the cognitive performances of ALS patients to describe the relationship between motor and cognitive dysfunction, according to site and side of disease onset.
Materials: Six-hundred and nine ALS patients underwent a comprehensive neuropsychological evaluation at diagnosis in Turin ALS Centre (MMSE, FAB, TMT A-B, Digit Span Forward and Backward, Letter Fluency, Category Fluency, RAVLT, BSRT, ROCFT, WCST, CPM47). Demographic and clinical data at the time of onset were collected too, encompassing side and site of onset, hand-dominance, age at onset, years of education and gender.

Methods:
Z- scores of neuropsychological tests were calculated and adjusted for age, gender and education. Cognitive performances of patients, grouped by side and site of onset, as well as clinical and demographic data, were statistically compared using $\chi2$ test, Kruskall-Wallis test and Mann Whitney U test.

Results:
Bulbar patients and bilateral spinal onset patients (Sbil) showed not significantly different performances across all neuropsychological tests, but lower cognitive performances in most of them, when compared to patients with lateralized onset (right-side spinal onset, Sri and left-side spinal onset, Sle). Left-side spinal onset patients (Sle) performed significantly better than right-side (Sri) at verbal memory tasks (RAVLT-dr, p=0.022 and BSRT-dr, p=0.033). When compared to bulbar patients, Sri patients performed better at Digit Span Forward (p=0.035) and Backward (p=0.009) and at visuospatial tasks (ROCFT-copy, p=0.007), while Sle patients performed better at verbal memory tasks (RAVLT-dr, p=0.030 and BRST-im, p=0.004).
Discussion: Spinal patients with symmetric motor impairment (Sbil) showed significant worse cognitive performance than those with lateralized damage and appeared to have a cognitive dysfunction similar to bulbar patients. Moreover, we found a strong lateralisation in neuropsychological tests for asymmetric spinal onset patients, reflecting hemispheric functional lateralization of language and visuospatial abilities. Specifically, Sle's left hemisphere relative sparing could justify their better performance at verbal memory tasks, while Sri's right hemisphere relative sparing could explain their better performances at visuospatial abilities and working memory tasks.

Conclusions:
Our findings point out that cognitive features of ALS patients are related to the lateralization of motor impairment, suggesting a possible disease spreading or parallel degeneration of highly interconnected frontal and precentral neurons (1). This is keeping with previous studies which found a direct relationship between lateralized motor and cognitive features (2,3).

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COG-09: Cognitive Reserve and Regional Brain Volume in Amyotrophic Lateral Sclerosis

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Objective:
We investigated whether cognitive reserve measured by education and premorbid IQ allows amyotrophic lateral sclerosis patients to compensate for regional brain volume loss.

Methods:
This was a cross-sectional study. We recruited sixty patients with amyotrophic lateral sclerosis from two specialist out-patient clinics. All participants underwent neuropsychological assessment; the outcomes were standardized z-scores reflecting verbal fluency, executive functions (shifting, planning, working memory), verbal memory and visuo-constructive ability. The predictor was regional brain volume. The moderating proxies of cognitive reserve were premorbid IQ (estimated by recognition-based vocabulary) and educational years. We hypothesized that higher cognitive reserve would correlate with better performance on a cognitive test battery, and tested this hypothesis with Bayesian analysis of covariance.

Results:
The analyses provided moderate to very strong evidence in favor of our hypothesis with regard to verbal fluency functions, working memory, verbal learning and recognition, and visuo-constructive ability (all BF01>3): higher cognitive reserve was associated with a mild increase in performance. For shifting and planning ability, the evidence was anecdotal (all BF01 between 1 and 3).

Conclusions:
These results indicate that cognitive reserve moderates the effect of brain morphology on cognition in ALS. Patients draw small but meaningful benefits from higher reserve, preserving fluency, memory and visuo-constructive functions. Executive functions presented a dissociation: verbally assessed functions benefitted from cognitive reserve, non-verbally assessed functions did not. This motivates future research into cognitive reserve in ALS and practical implications, such as strengthening reserve to delay decline.

COG-10: Cognitive Reserve in Amyotrophic Lateral Sclerosis (ALS): A population-based longitudinal study.

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Background:
Amyotrophic lateral sclerosis (ALS) is often associated with cognitive impairment. When cognitive changes emerge, and the extent of decline remains unclear. It is also unclear why some patients experience cognitive decline while others do not. In Alzheimer's, Parkinson's and Huntington's disease cognitive reserve (CR) has been used to account for why some individuals remain cognitively normal despite extensive neuropathology. CR theory posits that environmental enrichment through education, complex occupations and stimulating leisure activity facilitate compensatory mechanisms in response to neuropathology. Given the significant heterogeneity of ALS cognitive phenotypes, CR may play a protective role in offsetting cognitive impairment.

Aims:
This study examined the relationship between CR and longitudinal change in cognition in an Irish ALS cohort.

Methods:
Longitudinal neuropsychological assessment was carried out on 189 patients over 16 months using the Edinburgh Cognitive and Behavioural ALS Screen (ECAS) and a neuropsychological battery of tests. The neuropsychological assessment included tests of executive functioning, language, verbal fluency, social cognition and memory. CR was measured by combining education, occupation and physical activity data. Cox survival models were fit, with age of onset, diagnostic delay, site of onset and C9orf72 status as predictors of survival. Longitudinal mixed effects models were fit to examine the association between CR and cognitive performance over time. The longitudinal and time-to-event models were then combined to create joint models. The joint models examined the associations between CR, performance at baseline and decline over time while controlling for non-random drop-out.

Results:
Cognitive reserve was a significant predictor of baseline neuropsychological performance, with high CR patients performing better than those with medium or low CR. Better cognitive performance by high CR individuals was maintained longitudinally for ECAS, social cognition, executive functioning and language scores. Patients displayed little cognitive decline over the course of the study, despite controlling for non-random drop-out. This suggests that cognitive impairment tends to emerge early in the disease, with little decline thereafter.

Conclusions:
These findings suggest that cognitive reserve plays a role in the presentation of cognitive impairment at diagnosis, particularly in respect to ALS-specific deficits. CR was not protective against cognitive decline; however future research is needed with larger sample sizes and over longer time periods.

COG-11: Development of 'Coping And Living well with MND' (CALM), an online intervention to improve emotional wellbeing among people with MND and their caregivers

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Background:
We urgently need to develop psychological interventions for people with MND to improve emotional wellbeing and quality of life. Our study aimed to develop an online intervention (CALM) to improve emotional wellbeing among people with MND and their caregivers. We used the Person Based Approach (PBA) to ensure the intervention was acceptable, engaging and relevant to our target users by drawing on iterative qualitative research.

Methods:
We conducted two studies to understand our target users' perspectives on emotional wellbeing and psychological support. Study 1 was an in-depth qualitative interview study. 25 people with MND and 10 caregivers were interviewed about wellbeing and distress, data was analysed thematically. Study 2 was a systematic review of experiences of using psychotherapeutic and educational interventions to reduce distress among people with neurodegenerative diseases. Searches were conducted in 6 electronic databases. Qualitative data about participants' experiences of using interventions was extracted and thematically synthesized. Study findings and theoretical frameworks for psychological wellbeing during serious illness informed the development of CALM.

Results:

Findings from Study 1 highlighted key triggers of emotional distress (losing function and ability, having a threatened future, keeping up with multiple and constant changes, experiencing lack of support) and strategies to improve wellbeing (finding hope and positivity, exerting some control, being kinder to oneself, experiencing support). Based on these findings, CALM has sections on adjusting to changes, dealing with worries and stress, and building positivity and meaning. Key insights from Study 2 were that the intervention needed to accommodate various levels of disability, different coping preferences, and not be a burden for users. We designed CALM such that it was simple and easy to navigate, offered choice of techniques to use, and did not have a fixed set of sessions or homework to complete. Additionally, the language used was positive and empowering, not too confronting about death or the future. Guided by theory and these findings, we included advice and techniques from mindfulness-based approaches and compassion-focused therapy.

Discussion:
We used the PBA to understand the specific needs of people with MND and their caregivers and developed an online intervention tailored to their needs. We will continue to use in-depth qualitative studies to explore the acceptability, engagement and usability of CALM.

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COG-12: Preliminary Qualitative Analysis Of Focus Group Interviews Accessing Amyotrophic Lateral Sclerosis Caregivers Experiences Of Group-Based Interventions

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Live Poster Session B, December 10, 2020, 5:10 PM - 5:50 PM

Background:
Caregivers of patients with Amyotrophic Lateral Sclerosis (ALS) are often non-paid immediate family members who have a primary support role in patients' daily lives. However, caregiving brings challenges, and caregivers can have their own mental health needs that may go unmet (1). There has been an increased awareness of the need for practical and psychosocial supportive interventions for ALS caregivers, due to the complex nature of ALS (2). This study examines preliminary qualitive findings, resulting from focus groups that are part of an ongoing randomized controlled trial (RCT) with caregivers. Participants engaged in one of two group-based interventions, namely 'Mindfulness-Based Stress Reduction' (MBSR) and 'Building Better Caregivers' (BBC).

Method:
The first round of group based interventions was delivered in Beaumont Hospital, Dublin, Ireland. Due to Covid-19 restrictions, the second round of group based interventions was delivered on the online platform, Microsoft Teams. Qualitative data was gathered from five semi-structured focus group interviews with participants who completed the group-based interventions either in-person or online (N = 12). The semi-structured focus group interview addressed participants' attitudes toward the intervention programmes, suggested amendments, and ideas for general caregiver supports. The focus groups were delivered either over the telephone, online, or in-person. Two focus groups took place immediately following group completion. The remainder took place up to two months following group completion. One-to-one phone feedback with non-completers (N = 2), alongside informal feedback from non-completers is also discussed.

Results:
Themes arising from the analysis include Feasibility of the Group, Structure of the Programmes, Practical Issues, Support from Other Participants, The Impact of Group Facilitators, and Insights from Group Non-completers.

Conclusion:
Development and application of effective interventions for ALS caregivers requires detailed understanding of their complex needs, which can be further discovered by extensive thematic analysis.

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COG-13: International evaluation of current practices in cognitive assessment for amyotrophic lateral sclerosis

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Background:
Approximately 50% of people with amyotrophic lateral sclerosis (pwALS) will experience changes in cognition and/or behaviour, with 15% meeting the criteria for frontotemporal dementia (FTD), which is typically the behavioural variant (1, 2). The remaining 35% have milder, more specific changes, including impairments in executive functioning, language, social cognition, and apathy (3). In the UK, the assessment of cognition and behaviour has been widely implemented in ALS clinics, improving the quality of routine clinical care (4). Although cognitive and behavioural assessment is relatively well-established in the UK, there is a lack of knowledge on how these assessments are used in ALS clinics outside of the UK.

Objectives:
To investigate current practices in assessing cognitive and behavioural changes in international ALS clinics.

Methods:
In May-June 2019, directors, coordinators and/or principal investigators of international ALS clinics were invited via email to participate in an online survey about their current practices of using cognitive assessments in ALS. Survey questions explored: when and how assessments are undertaken, factors determining whether or not an assessment is carried out, and how the results affect clinical care and practice.

Results:

80/196 responded to the survey from 25 countries. 90% reported to assess patients for cognitive changes, with the Edinburgh Cognitive and Behavioural ALS Screen (ECAS) being the most popular test used, followed by the ALS Cognitive Behavioral Screen (ALS-CBS). These assessments usually took place in clinic and were often administered by a Clinical Neuropsychologist or Neurologist. 72% agreed that patients who are screened for cognitive change have better clinical care. However only 12.7% reported to assess all patients, and only 1/3 assessed patients more than once. When asked what the results were used for, respondents selected the following optional responses: inform and educate patients and carers about their difficulties (93.8%), inform management/intervention (87.5%), refer to other services (64.1%), and research (62.5%).

Thematic analysis of open-ended responses revealed that cognitive assessments: identify and validate changes in cognition and behaviour, aid understanding of the clinical impact of the disease, inform clinical care, and infer patient's decision-making abilities. Circumstances such as the clinical routine and logistical barriers affected when and which patients were offered cognitive assessments.

Conclusions:
Cognitive assessments have been implemented in ALS clinics around the world. Screening for cognitive and behavioural changes has benefits for clinicians, patients and carers, and is viewed as having a positive impact on clinical care.

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COG-14: Progression of cognitive and behavioural disturbances in motor neuron diseases assessed using standard and computer-based batteries

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Live Poster Session B, December 10, 2020, 5:10 PM - 5:50 PM

Background:
Detecting and monitoring cognitive and behavioural deficits in motor neuron diseases (MND) is critical due to their considerable clinical impact (1). The literature that so far has investigated the cognitive deficit progression of MND reported heterogeneous neuropsychological findings (2-5). In this scenario, computer-based batteries may play an important role.

Objectives:
To investigate the progression of cognitive and behavioural deficits in MND patients using both standard and computer-based neuropsychological batteries.

Methods:
This is a retrospective study on 74 MND patients (52 amyotrophic lateral sclerosis [ALS], 12 primary lateral sclerosis [PLS], and 10 progressive muscular atrophy [PMA]) who were followed up for 12 months and underwent up to three cognitive/behavioural assessments, 6 months apart, including standard and/or computerised based (the Test of Attentional Performance [TAP]) batteries. Behavioural/cognitive changes were investigated over time using generalized linear model for longitudinal data accounting for time and revised-ALS Functional Rating Scale.

Results:
Over 12 months, ALS patients showed a global cognitive decline (Mini Mental State Examination) at the standard battery and reduced performance in the alertness, sustained and divided attention, go/nogo, crossmodal and incompatibility TAP tasks. ALS patients did not show significant behavioural abnormalities over time. No cognitive and behavioural changes were found in PLS and PMA cases.

Discussion:
Computer-based neuropsychological evaluations, more than a standard cognitive assessment, are able to identify subtle cognitive changes in ALS, unique to this condition. This study highlights the need of specific, accurate and well-tolerated tools for the monitoring of cognitive deficits in MND.

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COG-15: Are psychiatric symptoms and disorders associated with cognitive and behavioural abnormalities in ALS?

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Live Poster Session C, December 11, 2020, 12:05 PM - 12:50 PM

Background:
Cognitive and behavioural impairment is common in amyotrophic lateral sclerosis (ALS), characterised by deficits in language, executive functioning, verbal fluency and apathy (1). However, memory dysfunction, disinhibition, loss of sympathy and/or empathy, perseveration and changes in food preference may also occur. These neuropsychological symptoms are present in up to 50% of patients, with ~10-15% meeting the criteria for frontotemporal dementia (ALS-FTD; 2). Higher rates of psychiatric disorders in ALS family members have also been reported (3,4,5).

Objectives:
To determine whether the presence of psychiatric symptoms in ALS kindreds is associated with cognitive and behavioural impairment in ALS patients.

Methods:
ALS patients and their first and second-degree family members were recruited in Scotland and Ireland. A comprehensive series of self-report questionnaires assessing a range of psychiatric symptoms were completed by the ALS kindreds. Current and lifetime, clinical and subclinical symptoms of mood (depression, mania), neurotic (anxiety, obsessive compulsive disorders), psychosis, suicidal thoughts, autism and attention deficit hyperactivity disorder (ADHD) were assessed. Cognition and behaviour in ALS patients were assessed using the Edinburgh Cognitive and Behavioural ALS screen (ECAS).

Results:
In total, 125 ALS patients with ECAS and personal or family psychiatric data were included. For ALS patients, current depression and anxiety was associated with poor language (exp(b)=0.619, p=0.017 and exp(b)=0.531, p=0.030). Current initiation apathy and impulsivity was associated with poorer memory (exp(b)=0.981, p=0.043 and exp(b)=0.981, p=0.044) and ALS non-specific scores (exp(b)=0.987, p=0.009 and exp(b)=0.986, p=0.006). An episode of mania or psychosis across the lifespan was associated with hyperorality (OR=8.667, 95%CI=2.065 to 37.370, p=0.003 and OR=6.800, 95%CI=1.482 to 30.145, p=0.011).

For ALS family member, higher impulsivity symptoms were associated with poor memory (exp(b)=0.98, p=0.02) and ALS non-specific (memory and visuospatial combined) scores (exp(b)=0.98, p=0.03) in ALS patients. However, several associations between family psychiatric symptoms and better ALS patient cognitive functioning were also shown. Higher current anxiety symptoms were associated with apathy (OR=1.40, 95%CI=1.07-1.97, p=0.03); autism with disinhibition (OR=1.29, 95%CI=1.06-1.67, p=0.02); and ADHD with hyperorality (OR=3.49, 95%CI=1.59-10.76, p=0.01).

Discussion:
Current psychiatric symptoms in ALS kindreds (depression, anxiety, impulsivity) are associated with poorer cognition in ALS patients, while lifetime psychiatric symptoms and disorders (anxiety, mania, ADHD, autism) showed more associations with behavioural features in patients. These results suggest psychiatric symptoms and disorders may contribute to the spectrum of frontotemporal symptoms in ALS, indicating a possible pleiotropy.

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COG-16: Development and validation of the Korean version of Edinburgh cognitive and behavioral amyotrophic lateral sclerosis screen (ECAS)

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Live Poster Session C, December 11, 2020, 12:05 PM - 12:50 PM

Background and objective:
Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disease of motor neurons with the incidence rate of about 2 per 100,000. It has been reported that approximately 30-50% of ALS patients show symptoms of cognitive impairment. Evaluation of cognitive and behavioral impairment is important as they are associated with shorter survival and increased caregiver burden. However, traditional cognitive evaluation tools have limitations because of the motor challenges and complexity of the cognitive impairment presented in ALS patients. Edinburgh Cognitive and behavioral ALS Screen (ECAS) is a brief multi-domain assessment tool, specifically designed for ALS patients. In this study, we aims to develop a Korean version of ECAS (K-ECAS) and evaluate its validity in Korean ALS patients.

Methods:
We plan to recruit 30 patients and 30 age- and education-matched controls, who visit Seoul National University Hospital and Seoul National University Boramae Hospital. Patients should be aged 40 or older, diagnosed as ALS according to El Escorial criteria, and native Korean speakers. Those who had history of psychologic other neurologic disease are excluded. Control group was chosen from healthy carer of the patient, matched by age and educational status. To test the construct validity, the patients will be tested with Montreal Cognitive Assessment (MoCA), Frontal assessment battery (FAB) and Center for Neurologic Study-Liability Scale (CNS-LS).

Results:
ECAS was translated into Korean from the original version of ECAS according to cross-cultural adaptation guideline, using the standard multistep forward-backward method. Total and subdimension scores of the K-ECAS will be compared between patients and controls, and the correlation with other measures of cognitive evaluation will be conducted.

Discussion:
This study is the first to develop and validate a Korean version of ECAS. With validation of K-ECAS, we hope to screen cognitive and behavioral impairments more effectively in our ALS patients.

COG-17: Early Evolution and Evaluation of a New MND Clinical Neuropsychology Service

Miss Rosie Lesley[1], Dr Emily Mayberry[1], Miss Theresa Walsh[2], Mr Lee Tuddenham[2], Mrs Helen Wollff[1], Mrs Mbombe Kazoka[2], Rosie Whitehead[1], Dr Pete Walpole[1], Dr Claire Tooth[1], Dr Esther Hobson[2], Dr Channa Hewamadduma[2], Dr Thomas Jenkins[2], Professor Christopher McDermott[2], Professor Dame Pamela Shaw[2]
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Live Poster Session C, December 11, 2020, 12:05 PM - 12:50 PM

Background:
Motor neuron disease (MND) was historically believed to affect the body but not the mind. More recently, research has clearly shown that approximately half of people with MND experience cognitive changes. For some, the changes are very subtle, but for roughly ten percent of people with MND, cognition and behaviour are profoundly affected.

Establishment of the Sheffield MND Clinical Neuropsychology Service:
Originally, the primary aim of the Sheffield MND Clinical Neuropsychology service was to improve care planning for people with MND through early identification of cognitive change and prompt access to further neuropsychology assessment and advice. Therefore, the initial focus was on establishing and improving the cognitive screening pathway.

Evaluation and User Feedback:
Quantitative and qualitative feedback has been received from 18 patients who had experience of completing the cognitive screening and 25 healthcare professionals who were involved in the care of patients who had completed screening. Patient satisfaction data has also been received from 18 patients seen through the MND neuropsychology pathway. Several changes to the service have been made in response to this feedback.

Changes in Service Provision:
Additional psychological needs have been identified, and the service has been adjusted to better meet these needs. In particular, a need for more person-centred and holistic assessment (including consideration of mood, anxiety, social factors, and carer support needs) has been identified, as well as a need for emotional support.
All aspects of the service are now informed by Acceptance and Commitment Therapy (ACT), which aims to help people clarify what matters most to them, and find ways of doing more of these valued activities in the face of practical and emotional obstacles.
Therefore, the MND Clinical Neuropsychology service aims to facilitate early conversations about what matters to each person with MND and their families so that this can inform all aspects of assessment and ongoing care.

Challenges and Future Directions:
We have encountered a number of challenges during the development of the service, including ensuring people with MND understand the screening process and feel they have a choice about completing screening. We have also had to think about when the best time to offer screening is, how to do this in a holistic way, what to do when people decline further assessment/intervention, and how to overcome issues around accessibility and fatigue. More widely, we have had to consider how to sensitively and effectively share screening results with people with MND and other healthcare professionals.
Future directions for the service include increasing patient choice and accessibility through the use of technology, offering more intervention work, refining and evaluating the MND neuropsychology pathway, and sharing learning and increasing collaboration with other MND care centres.

Thanks to the MNDA for their support.

COG-18: Social Cognition deficits in Amyotrophic Lateral Sclerosis: a population based study

Dr Francesca Palumbo[1], Dr Barbara Iazzolino[1], Dr Laura Peotta[1], Dr Antonio Canosa[1,2], Dr Rosario Vasta[1], Dr Umberto Manera[1], Dr Cristina Moglia[1,2], Prof. Adriano Chiò[1,2], Prof. Andrea Calvo[1,2]

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Introduction:
Social Cognition (SC), in particular Emotion Recognition (ER) and Theory of Mind (ToM), has been intensively studied in the last decades in several neurodegenerative diseases, including Amyotrophic Lateral Sclerosis (ALS). To date, most studies report an impairment both in ER an ToM in ALS patients and deficit in SC have been included in ALS-frontotemporal spectrum disorders revised diagnostic criteria (Strong, 2017). There is, however, remarkable heterogeneity in phenotypic characteristics of patients and neuropsychological tools across different studies.

Objective:
To assess Social Cognition (SC) profile in ALS patients at diagnosis. Secondly, to assess possible differences in SC abilities depending on cognitive profile, motor phenotype and degree of severity of motor impairment.

Methods:
We included fifty-two patients attending the ALS Center of Turin University Hospital between May 2019 and July 2020. All patients underwent clinical assessment by means of ALS Functional Rating Scale, Neuropsychological assessment and SC assessment. This was conducted through Ekman 60 faces test (ET), Reading Mind in Eyes task (RMET), Story-Based Empathy task-Emotion Attribution (SET-EA) and Intention Attribution (SET-IA). Both SET-IA and SET-EA were compared to a control condition of Causal Inference (SET-CI).

Results:
We subdivided patients into cognitive categories, according to Strong revised criteria (2017), as follows: 25 cognitively normal (ALS-CN), 13 with cognitive impairment (ALSci), 4 with behavioural impairment (ALSbi), 7 with cognitive and behavioural impairment (ALScbi), 3 with dementia. Not demented patients with some degree of cognitive and/or behavioural impairment (ALSci-bi-cbi) performances were significantly worse compared to CN patients in RMET (p 0.003), SET-IA (p<0.001) and SET-EA (p<0.001), as expected. However, also CN patients showed deficit in at least one SC test (10 out of 25). Specifically, 4 of them showed deficit in SET-EA. Moreover, multiple linear regression analysis showed significant correlation between SET-IA score and ALSFRS-total score (p 0.012). Finally, out of 4 patients with c9orf72 mutation, defined as CN at diagnosis, 3 showed selective deficit in ER.

Discussion:
Our findings support the hypothesis that ALS-CN patients may show impairment in all SC subcomponents since the early stages of disease. Moreover, we show a progressive related worsening of cognitive subcomponent of Theory of Mind (Intention Attribution) and severity of motor symptoms. Further studies are necessary to evaluate SC impairment pattern in ALS patients and its eventual imaging and genetic correlates both to delineate a more precise definition of cognitive profile and also to elaborate better care strategies taking into account eventual deficits in managing emotional material in such patients.

COG-19: Taking the Scenic Route: Discussion on Adapting a Group Intervention RCT for ALS Caregivers to Online in Response to COVID-19

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Live Poster Session C, December 11, 2020, 12:05 PM - 12:50 PM

Background:
Caregivers of people with ALS, commonly non-paid immediate family members, often take primary responsibility for the complex care needs of patients outside of the medical setting. Recent longitudinal findings suggest that in ALS patient-caregiver dyads, caregivers' experience of burden is associated with a higher baseline of anxiety and depression, which leads to a reduced QoL as the disease progresses. This study aims to evaluate the effectiveness of two types of low-intensity intervention groups for mild-to-moderate anxiety, depression, QoL and burden in caregivers of people with ALS, with a cohort of Irish caregivers. One full round of intervention groups was delivered in-person, prior to COVID-19 related restrictions being put in place in Ireland. As a result of the restrictions, in-person contact with caregivers was no longer possible. New solutions were developed to enable continued recruitment of participants and delivery of group interventions, despite the challenges presented by the pandemic.

Method:
The practical steps involved in adapting the study included; applying for ethical approval to make the relevant amendments, adapting the psychometrics for online delivery, embedding checks to ensure safety in relation to measures of distress, creation of an online information video, making contact with potential participants through various social media platforms, notifications and relevant staff contacts, engaging a platform for online group work and making approved adaptations to intervention formats to enable online delivery.

Discussion:
Moving the RCT from the hospital setting to online has presented many challenges but facilitated new possibilities, such as allowing caregivers in areas geographically distant from the location of the study to attend group. Delivering online therapy groups presents therapeutic issues in terms of managing boundaries and creating a containing space for sharing experiences and feelings. Particular attention was given to boundary-setting and participant self-support, as well as incorporating a level of flexibility that enabled caregivers to attend group in the context of multiple competing demands. Facilitating group cohesiveness, an important factor in any therapeutic group, also required special consideration in adapting the intervention to online.

COG-20: The impact of COVID-19 on individuals with motor neuron disease and their caregivers

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Live Poster Session C, December 11, 2020, 12:05 PM - 12:50 PM

Background:
Patients with ALS demonstrate anxiety, depression, and hopelessness in higher levels than the general population (Felgoise et al., 2010). Caregivers often carry substantial psychological and physical burdens (Pagnini et al., 2010). Researchers have not studied the impact of the COVID-19 pandemic on MND patients and their caregivers, although many are living under conditions that limit their mobility and social interactions beyond those already posed by their underlying illness.

Objective:
To evaluate and describe the psychological impact of the COVID-19 pandemic on patients with MND and their caregivers.

Methods:
Patients with MND and their caregivers were recruited by email in collaboration with the ALS Association Greater Philadelphia Chapter. Subjects provided e-consent and completed a one-time survey via REDCap. The survey was comprised of questions from the ALS Functional Rating Scale, ALS Specific Quality of Life Short Form, Zarit Burden Interview, and novel qualitative and quantitative research questions to assess the experience of subjects during the COVID-19 pandemic.

Results:
Responses were received from 87 patients and 59 caregivers. 50.6% of patients and 69.0% of caregivers were women. Patients had an average age of 65.5(SD=10.7) and median time since symptom onset of 43 months (range:4-291). Caregivers had an average age of 66.3(SD=57.9). ALS was the most commonly reported type of MND (87.4% of patients). Of 122 respondents, only 1 tested positive for COVID-19 and 1 reported having symptoms. When asked how the pandemic has changed their life, 4(3.3%) indicated much worse, 77(63.1%) somewhat worse, 35(28.7%) unchanged, and 6(4.9%) somewhat better. 47.7% of patients reported that the pandemic is not affecting them physically. Main worries included contracting or having a loved one contract COVID-19(26.9%), MND-related worries (20.4%, predominantly patients), and taking care of others (15.7%, predominantly caregivers). When asked what has changed during the pandemic, patients reported less freedom (25.4%), MND-related concerns (23.8%), and social isolation (17.5%), while caregivers reported less outside or recreational activities (27.1%), and social isolation (14.6%). 67.8% of subjects do not think their health has been affected by the pandemic. 73.9% reported changing their behavior since the beginning of the pandemic.

Discussion:
Patients reported worrying more about MND-specific symptoms than contracting the virus, while caregivers expressed a strong concern about the virus, together with the need to take care of others. The loss of freedom due to restricted movement and social isolation are the main changes that have been reported by patients. Mental health reduction was sometimes cited, together with a general feeling of being more dependent. Additional analysis is needed to further explore the results from this study.

COG-21: The Interplay Among Education, Brain Metabolism, and Cognitive Impairment in Amyotrophic Lateral Sclerosis Supports a Role of Cognitive Reserve

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Aim:
To test the Cognitive Reserve (CR) hypothesis in ALS.

Methods:
We enrolled 111 ALS patients. We used years of education as CR proxy, 18F-FDG-PET to assess brain lesion load, and the Edinburgh Cognitive and Behavioural ALS Screen (ECAS) to measure cognitive functioning. Education was regressed out against whole brain metabolism, including age, sex, site of onset, ALSFRS-R total score, and ECAS score as covariates. Since the presence of the hexanucleotide repeat expansion of C9orf72 can impact on brain metabolism, we performed a sensitivity analysis with the same procedure on the patients for whom the genetic analysis resulted negative for C9orf72, SOD1, TARDBP, and FUS mutations. Metabolic clusters showing a significant correlation were used as seed regions in an interregional correlation analysis (IRCA) to identify regions whose metabolism was correlated with that of the seed clusters. We also performed the IRCA in a control group (n=40) using the same seed regions.

Results:
In the ALS group we found a negative correlation between brain metabolism and education in right anterior cingulate and bilateral medial frontal gyrus. In the IRCA in the ALS group the metabolism of the medial frontal cluster positively correlated with that of frontotemporal regions (right>left), bilateral caudate nuclei, and right insula, and negatively correlated with that of corticospinal tracts, cerebellum and pons. In the control group the IRCA showed very limited correlations as compared to the ALS group. Above all, the negative correlation between the medial frontal cluster and the cerebellum was not found in the control group.

Conclusion:
Our results are in agreement with the CR hypothesis, since higher education is associated with lower metabolism in frontal regions, independently from the level of cognitive decline, in ALS patients. The negative correlation between the medial frontal cluster and the cerebellum that we found in the IRCA only in the ALS group might reflect cerebellar involvement in compensation to frontal cognitive damage.