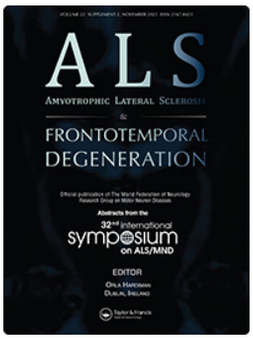

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Theme 12 - CLINICAL MANAGEMENT, SUPPORT AND INFORMATION

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THEME 12 CLINICAL MANAGEMENT, SUPPORT AND INFORMATION

CMS-01 Overnight oximetry detects respiratory dysfunction earlier in ALS

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Background/Introduction: Non-invasive ventilation (NIV) improves survival time in people living with ALS by seven months on average (1). Recent studies suggest that initiation of NIV at earlier respiratory parameters is associated with improved survival (2). Our study objective was to determine if overnight nocturnal oximetry (ONO) diagnosed respiratory insufficiency before in-office spirometry respiratory parameters (RP).
Methods: This study was a retrospective review of patients in a multidisciplinary ALS clinic at a major academic center. 200 de-identified patient charts with active NIV orders were randomly selected for review. Parameters collected were: sex, age at diagnosis, current age, site of disease onset, date of first NIV order, spirometry and ALS functional rating scale (ALSFRS) on day of NIV order and reason for NIV order (abnormal ONO versus in-office spirometry meeting Medicare guidelines).
Results: 175 patients had complete records for review. Reason for NIV order was abnormal ONO for 84 patients (group ONO) and abnormal in-office spirometry for 91 patients (group RP). Age of onset, distribution of onset site, and FRS were the same between groups. On the day of NIV order, the ONO group had significantly better FVC (70% vs 51.9%, $p < 0.001$) and MIP (−57.2 vs −32.8 cm H_2O, $p < 0.001$) when compared to RP group.
Discussion: Our retrospective review showed that respiratory dysfunction from motor neuron disease was detected earlier with overnight oximetry than in-office spirometry. This is a significant finding especially as the COVID-19 pandemic continues and there are significant safety concerns with performing aerosolizing procedures like spirometry and increasing telehealth visits. Further study is needed to determine how earlier initiation of NIV affects quality of life and overall survival in patients with ALS.

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References

1. Bourke SC, Tomlinson M, Williams TL, et al. Effects of non-invasive ventilation on survival and quality of life in patients with amyotrophic lateral sclerosis: a randomised controlled trial. Lancet Neurol. 2006;5(2):140–7.
2. Khamankar N, Coan G, Weaver B, et al. Associative increases in amyotrophic lateral sclerosis survival duration with non-invasive ventilation initiation and usage protocols. Front Neurol. 2018;9:578. doi: 10.3389/fneur.2018.00578.

CMS-02 Tongue measurements and oral intake level in patients with ALS

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Background: Tongue pressure and endurance are impaired by muscle weakness and atrophy in patients with ALS (1). Therefore, adaptations in the management of liquid and food oral intake could be necessary to maintain the efficiency and safety of swallowing in these patients.
Objectives: to correlate maximum tongue isometric pressure (MTIP), tongue isometric endurance (TIE), and tongue thickness (TT) with liquid and food oral intake in patients with ALS.
Methods: Twenty-one patients (mean age 60.21 ± 13.21 years, 61.9% male) with ALS (38.1% bulbar type; 61.9% spinal type) were included for MTIP and TIE assessments by the Iowa Oral Performance Instrument (IOPI) (1), TT assessment by ultrasonography (US) (2) and the level of oral intake by the Functional Oral Intake Scale (FOIS). In this study, the seven-level FOIS were divided into two categories: <7 and 7.
Results: Patients with a FOIS score <7 had a significantly lower MTIP ($p = 0.01$) compared to patients with a FOIS score of 7. There was no association between TIE or TT and FOIS.
Discussion: In this study, MTIP was lower in patients with impaired FOIS (<7 level). This finding reinforces the relevance of tongue assessment since the early diagnostic investigations, with the purpose of monitoring the progression of the disease and minimizing the negative impact on oral intake, nutritional status, and the quality of life. Quantitative measurements of the tongue, especially MTIP, could also seem to be a possible predictor of fatigue and functional oral intake impairment, which represents valuable guidance for clinical decision-making on dysphagia and eating management and rehabilitation (3).

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References

1. Hiraoka A, Yoshikawa M, Nakamori M, et al. Maximum tongue pressure is associated with swallowing dysfunction in ALS patients. Dysphagia 2017. 32(4):542–7.
2. Nakamori M, Hosomi N, Takaki S, et al. Tongue thickness evaluation can predict swallowing function in amyotrophic lateral sclerosis patients. Clin Neurophysiol. 2016;127(2):1669–74.
3. Pizzorni N, Ginocchio D, Bianchi F, et al. Association between maximum tongue pressure and swallowing safety and efficacy in amyotrophic lateral sclerosis. Neurogastroenterol Motil. 2020;32(8):e13859.

CMS-03 ThinkALS: a user-friendly and comprehensive ALS diagnosis and referral tool for general neurologists

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Background: ALS diagnosis is complex and requires a thorough clinical assessment and focused diagnostic work up. In the U.S, the average time from weakness onset to diagnosis confirmation is 11.5–15 months, during which a patient sees average of 3–4 medical providers prior to diagnosis confirmation (1,2). Reducing the time to initial ALS neurologist consultation will greatly reduce diagnostic timelines. Based on several factors, the ALS Association's spring 2020 multi-stakeholder Roundtable meeting identified, that US general neurologists would be our first key target population for clinician education and awareness initiatives.
Objectives: To increase ALS education and awareness among general neurologists in the U.S. The aim is to promote early ALS suspicion and improve efficiency of early referrals to ALS multidisciplinary clinics for diagnosis confirmation and treatment initiation.
Methods: A multidisciplinary initiative led by The ALS Association, created a national Time-to-Diagnosis working group, which comprised of 18 members including experienced ALS neurologists, patients/caregivers, industry representatives, and ALS Association leaders. From 2020-present date, the group meets at frequent intervals, has conducted thorough literature reviews, administered nation-wide surveys to ALS and non-ALS clinicians, and patients/caregivers to gather data on the diagnostic barriers, understand current referral processes and timelines, brainstorms solutions and creates clinician educational interventions including diagnostic guides and referral tools.
Results: Through an iterative process the working group developed a simple and comprehensive thinkALS tool, which serves as a diagnostic guide and a referral tool for general neurologists. The tool provides information on clinical characteristics and high yield examination findings for ALS, and in one place, provides resources and search tools for the nearest multidisciplinary ALS clinic and guidance on how to write ALS referrals that would expedite triage and consultation in most specialized ALS care centers. Educational initiatives in the UK and Australia led to the development of the ALS Red flag tool targeting primary care physicians to which we based thinkALS (3). The thinkALS tool has been presented to and revised to its current final form, based on formal surveys and informal group discussion feedback from clinician teams and leaders at the AAN, AANEM, NEALS, and the UK and Australia MND Association. A dedicated website, www.thinkals.org, is being created to house the tool and other clinician educational resources focused on early diagnosis.
Discussion: The thinkALS tool and website will support general neurology clinicians to increase their confidence to suspect ALS early and lead expeditious referrals to dedicated multidisciplinary ALS clinics.

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The Time-to-Diagnosis working group members, AAN neuromuscular leadership, members and leadership of AANEM, and NEALS.

References

1. Paganoni S, et al. ALSFTD. 2014;15:5–6.
2. Benatar M. ALSFTD. 2019;20;5–6.
3. www.mndassociation.org/professionals/management-of-mnd/management-by-specific-professions/information-for-gps/red-flag-diagnosis-tool/

CMS-04 Veterans with ALS and suicidal ideation: a community created intervention

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Background: An ALS diagnosis presents many physical, social and psychological challenges. A recent piece, "Suicide among veterans with amyotrophic lateral sclerosis" ("Suicide"), highlights that an ALS diagnosis leads to an increased risk of mental health illness, including depression, suicidal ideation and death by suicide. This is particularly true for veterans who are at a higher risk for suicidal ideation and death by suicide. (Lund) I AM ALS is a community-led US non-profit that revolutionizes ALS Advocacy. Community members self-organize themselves into community teams, such as the Veterans Affairs Team. This team meets once a week to raise awareness for veteran-specific ALS issues and connect veterans with resources to improve their quality of ALS care. Upon reading "Suicide," the Veterans Affairs Community Team decided to create a program that would raise awareness around mental health issues and resouces for veterans with ALS and decrease the occurrence of suicidual ideation and death by suicide.
Objectives: To understand, raise awareness and intervene in suicidal ideation and death by suicide.

Methods: I Am ALS Veterans Affairs Team conducted a literature review, which informed a three-stage social media campaign to destigmatize mental health, reframe ALS as something people are living with not dying from and connect people to medical, social and advocacy resources.
Results: The Veterans Affairs Team has been working with veterans, caregivers and mental health professionals to identify triggers for mental health crises related to an ALS diagnosis that could lead to suicidal ideation. The team has worked together to create a multimedia social media campaign that aims to destigmatize mental health issues and offer support and resources to veterans and caregivers.
Discussion: An extraordinary amount of time within the ALS landscape is spent talking about the physical impact of ALS. This campaign is important as it raises awareness of the mental impact of ALS and connects people to medical and non-medical resources. The I AM ALS Veterans Affairs Team will continue to expand upon this initial campaign to further their goals of connecting veterans with ALS to the resources they need.

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We would like to thank the ALS patients and caregivers that were so open and willing to discuss this important issue.

Reference

1. Lund EM, Hostetter TA, Forster JE, et al. Suicide among veterans with amyotrophic lateral sclerosis. Muscle Nerve. 2021: 1–5.

CMS-05 A Clinical bulbar scale for ALS/MND (C-BAS): Preliminary validation

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Background: Comprehensive, clinically reliable bulbar assessment methods for use within MND/ALS clinics remain limited to date. Identification of bulbar onset accurately predicts the progression of motor neuron impairment, distinguishing from cervical, thoracic, and lumbosacral body regions (1). Critical to reliable quantification of bulbar symptoms is specifying features of upper motor neuron (UMN) and lower motor neuron (LMN) impairment in functional, clinically observable tasks. Longitudinal clinical measurement of cumulative bulbar functional tasks may aid identification of treatable factors influencing ALS disease progression (e.g. weight, respiration, speech intelligibility, speaking rate, vocal quality, communication effectiveness) (2). C-BAS is designed to standardize clinical bulbar function, identifying the broader goal of providing reliable data collection for clinical and research application. The C-BAS compilation of measures has been developed to identify and track progression of bulbar function over time, by quantifying speech and swallowing features, maintaining sensitivity to longitudinal change, and characterizing functional impacts.
Hypothesis: C-BAS will effectively identify early bulbar impairments, facilitate timely clinical interventions, accurate disease progression tracking, and clinical research trials.
Methods: The C-BAS represents a collection of objective and subjective clinical bulbar measures, designed to assess speech and swallowing impairments while tracking progression in ALS. Participants were measured on speech and swallowing tasks, UMN and LMN features, and self-reported effort, fatigue, and communication/swallowing effectiveness. Existing scales (e.g. ALSFRS (3), CNS-BFS (4)) were completed in parallel for comparisons.
Results: Preliminary data analysis indicates:
1. C-BAS item content validation (CVR >0.99).
2. Ongoing data collection, with analyses of ($n = 54$) participants to date. C-BAS scores range from 8 to 93 ($M = 38.56$, $SD = 24.1$); a trend in C-BAS scores is noted (i.e. bulbar higher ($M = 56.2$, $SD = 24.8$); spinal lower ($M = 36.0$, $SD = 23.8$)).
3. Strong significant correlations were identified with existing bulbar scales: (e.g. ALSFRS-R, CNS-BFS) with ALSFRS-R ($r = -.71$, $p < 0.001$), CNS-BFS ($r = .84$, $p < 0.001$).
Conclusions: C-BAS incorporates systematic data collection, involving a clinical assessment of speech and swallowing subsystems using a combination of subjective and objective clinical measures, which may determine overall impact of bulbar symptoms on personal and social activities. Preliminary evidence suggests a valid, reliable scale for objectively assessing bulbar motor function in patients with MND/ALS.

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References

1. Yorkston KM. The degenerative dysarthrias: a window into critical clinical and research issues. Folia Phoniat Logop IALP. 2007;59(3):107–17.
2. Traynor BJ, Alexander M, Corr B, et al. Effect of a multidisciplinary amyotrophic lateral sclerosis (ALS) clinic on ALS survival: a population-based study. 1996-2000. J Neurol Neurosurg Psychiatry. 2003;74(9):1250–61.
3. Gordon PH, Miller RG, Moore DH. ALSFRS-R. Amyotroph Lateral Scler Mot Neuron Disord. 2004;5(Suppl 1):90–3.
4. Smith RA, Macklin EA, Myers KJ, et al. Assessment of bulbar function in amyotrophic lateral sclerosis: validation of a self-report scale. Eur J Neurol. 2018;25(7):907–e66.

CMS-06 Applying a clinical algorithm on real-world Electronic Medical Record (EMR) data for patient risk stratification of undiagnosed amyotrophic lateral sclerosis (ALS)

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Background: ALS is a progressive neurodegenerative neuromuscular disease that is challenging to diagnose. Early diagnosis and referral to a specialized multidisciplinary centre may improve patient outcomes. No single test definitively confirms disease. As such, algorithmic tools to aid in diagnosis are needed. The introduction of novel computing tools that facilitate analysis of structured and unstructured electronic medical record (EMR) data using natural language processing has made the development and application of such tools in clinical practice feasible.
Objectives: We investigated the feasibility of developing and applying a clinical algorithm to real-world EMR data to aid in diagnosis of ALS.
Method: A clinical algorithm was developed and applied using the Advanced Computing Platform by Ensho Health. Our clinical algorithm was designed to screen structured and unstructured data for clinical and electromyography (EMG) findings of lower and upper motor neuron abnormalities in four spinal regions. The algorithm was applied to the de-identified electronic health records of 3372 patients at a single community neurologist practice in Ontario, Canada.
Results: 1332 patients (39%) had evidence of at least one clinical feature screened for by the algorithm and were categorized as Very High (12%), High (3%), Moderate (<1%) and Low (84%) Risk. Of the 160 (12%) of patients categorized as Very High Risk, 3 (2%) had a diagnosis of ALS (100% sensitivity), 7 (4%) had pending workups for ALS (80% sensitivity) and 9 (6%) had ALS ruled out in prior clinical investigations (61% specificity). Alternative explanatory diagnoses subsequently excluded 129 (91%) of the 141 Very High Risk patients without previous suspicion for ALS. Records of the remaining 12 (9%) patients were reviewed clinically. 11 (92%) had alternative explanations for the features incorporated in the algorithm and 1 (8%) likely had undiagnosed ALS but passed away before completing further investigation.
Discussion: Clinical algorithms can be applied to real-world EMR data to aid in diagnosis of ALS in a neurological community practice. Our initial algorithm weighed subjective findings too heavily and considered too few alternative diagnoses when narrowing results for clinical review, which we attribute to the observed false positive rate. More importantly the algorithm detected 1 patient with likely undiagnosed ALS as Very High Risk and had 100% sensitivity in categorizing all 3 known ALS patients in the practice as Very High Risk. A limitation of the practice setting was that complex neurological patients were being referred to a tertiary neurological centre for management, potentially limiting the number of undiagnosed Very High risk patients for subsequent investigation. Further refinements to the algorithm are planned as are expanding its application to more pilot sites including to multidisciplinary centres for referral evaluation.

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CMS-07 Impact of established respiratory home care protocols on the care of the motor neuron disease patients during the COVID-19 pandemic

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Background: For the last five years, established home respiratory care protocols have been used by the VCU Health ALS Clinic These protocols have increased non-invasive ventilation (NIV) compliance, decreased hospital admissions for respiratory issues, and fostered communication with patients with ALS (PALS), caregivers, the respiratory home care team, and the interdisciplinary staff in the ALS clinic.
Objective: To assess the utility of home respiratory protocols to enhance ALS telehealth visits and goals of care discussions for PALS not seen in person in the ALS clinic during the COVID-19 pandemic from 1 March 2020 to 31 March 2021.
Method: During March 2020 to March 2021, 163 patients were seen in the VCU Health ALS Clinic. Of these 163 patients, 55 were on NIV prior to March 2020. There were 53 patients new to the clinic, or were seen for a confirmation of diagnosis in the clinic. There were 29 patients who met criteria for initiation of NIV, mechanical insufflation-exsufflation device(cough assist), and suction machine. Each patient was assigned a home care respiratory therapist who had been trained on the VCU Health ALS Clinic respiratory protocols. Every patient was also encouraged to view a 25-min video that discussed respiratory care of the ALS patient, with demonstration of mouthpiece ventilation, nocturnal ventilation, cough assist, and suction machine usage. Every 6 weeks in the patient's home, the respiratory therapist captured the following data points: end tidal carbon dioxide level, NIV compliance, full download of NIV settings (including leak percentage), upright and supine forced vital capacity (FVC), oxygen saturations, and use of cough assist. Additional data included documentation of any observations of sialorrhea, bronchial secretions, falls, COVID 19 exposures, and patient or caregiver concerns. The clinic team reviewed all data points and worked with home care teams to correct leaks, increase NIV compliance (goal 8/24 h), and adjust NIV settings to address decline in respiratory function during disease progression. The home care teams also had alerts for an immediate clinic call if the patient's FVC approached 50% of normal. This was to facilitate discussion with the neurologist, dietitian, and respiratory therapist concerning placement of a feeding tube. An alert was also used for FVC approaching 30%, allowing goals of care discussion to take place to document end of life wishes or facilitate tracheostomy placement. In this time period, 28 patients died from complications of their disease.
Conclusion: During the COVID-19 pandemic, established respiratory home care protocols enhanced coordination of care for PALS in the VCU Health ALS Clinic. Quantifying the decline of respiratory function and disease progression in PALS, unable to be seen in person, enabled the providers to continue to deliver high quality patient-centric care in the telehealth environment.

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CMS-08 The association of passive smartphone mobility measures and communicative participation in ALS

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Background: Communication decline is a devastating consequence of ALS that impacts daily functioning and social connection. People with ALS (PALS) report limitations to their communicative participation (communication in daily life situations). Understanding potential factors associated with communicative participation, such as reduced engagement in life activities outside of the home, is key to monitoring and mitigating these restrictions. Analysis of Global Position System (GPS) data has been used to index mobility in PALS and provides the opportunity to quantify communicative participation opportunities outside of the home. Because it can be passively collected on smartphone platforms, this low-cost and low burden approach is ideal for overcoming the common barriers to clinical and research participation experienced by PALS.
Objective: To evaluate the utility of passive smartphone metrics to index and monitor communicative participation in ALS, this study explores the associations between measures of passive smartphone mobility data (GPS) and communication function.
Methods: Data were collected from 25 PALS during periods before and after COVID-19 pandemic restrictions. Multiple data streams were collected using the Beiwe smartphone research platform. Participants completed surveys – the Communicative Participation Item Bank Short Form (CPIB) and the Revised ALS Functional Rating Scale (ALSFRS-R) – and recorded themselves reading a passage aloud. Multi-week GPS data were analyzed across variables related to time and distance spent away from home. Statistical analyses included bivariate correlations between variables and Welch's *t*-tests to compare participants with versus without participation restrictions.
Results: The mean (SD) ALSFRS-R bulbar subscale scores was 10.61 (1.95) and ranged from 5 to 12 across all participants. Eleven participants reported communicative participation restrictions. CPIB scores were significantly correlated ($p < 0.001$) with bulbar measures, including the ALSFRS-R bulbar subscale and speaking rate. For GPS variables, time spent at home was inversely related to CPIB scores ($r = -0.49$, $p = 0.014$) and speaking rate ($r = -0.49$, $p = 0.018$). Time spent at home by participants with communicative participation restrictions (mean =18.4 h) was significantly higher than for participants without (mean =14.4 h) restrictions ($t(21.7) = 2.53$, $p < 0.05$). In addition, time spent at home was associated with the ALSFRS-R gross motor subscale ($r = -0.51$, $p = 0.017$). Variables indexing distance traveled were not significantly associated with communication and functional variables.
Discussion: Our preliminary findings indicate moderate associations between the time PALS spent at home and communicative participation, bulbar, and motor function. Further investigation of these relationships is warranted, as passive data collection to identify and monitor communication function is a promising approach for addressing communicative participation decline and supporting social connection in ALS.

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CMS-09 Improving online caregiver training for ALS and complex fragile patients using design-build in Italy

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Background: Accessibility to caregiver training for ALS and complex fragile patients became further complicated during the COVID-19 pandemic for all those who engage in caregiving.
Objectives: Our presentation examines how a patient led, multi-disciplinary Italian ALS association works to improve caregiver training access in Italy through a new online course.
Methods: The course has been developed from an innovative interdisciplinary design-build approach which integrates feedback from the Italian ALS community to develop caregiver resources (1). This new online course launch is Italy's first free caregiver training program available on demand 24 h a day, 7 days a week for ALS and complex fragile patients covering fourteen essential themes in a user-friendly format. Users are requested to complete a survey at the completion of the course.
Results: Since its launch in January 2021, preliminary data gathered illustrate high enrollment (120 users). We received 27 surveys from across the country with the following results: 26 users recommended the course to others and 1 user did not because they found it too advanced.
Discussion and conclusion: Additional reminders to users will be sent to obtain completed surveys. Although we achieved widespread enrollment across Italy, expansion of course offerings will continue while improving content based on feedback received from the clinical and caregiver field settings.

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Reference

1. Zicchieri AD, Conte SF, De Rossi N. CMS-27: using a design-build approach to improve international ALS research news access in Italy. Poster presented at the Motor Neurone Disease Association Virtual 31st International Symposium on ALS/MND, 9–11 December 2020; 2020.

CMS-10 Novel respiratory therapy combining expiratory muscle strength training and air stacking in patients with amyotrophic lateral sclerosis: videofluoroscopic findings in the upper airway

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Background: Considering the respiratory complications that arise in amyotrophic lateral sclerosis (ALS), the development of new therapies to improve bulbar and respiratory impairments must be required. In a clinical trial conducted by Plowman et al., expiratory strength training (EMST) promoted improvements in respiratory and bulbar function in patients with ALS (1). Among the most commonly used strategies in respiratory management is the periodic lung expansion by air stacking (AS) which decreases basal atelectasis, maintains compliance of the lungs and chest wall, and increases peak cough flow and cough effectiveness. Considering previous results, Dorça et al. (2) developed a novel respiratory therapy (RT) using a modified bag-mask ventilation system coupled to a manometer and a unidirectional valve with PEEP to perform EMST on the expiratory phase of AS. However, the clinical outcomes of this therapy remain unclear.
Objectives: To measure the acute effects of this therapy on the upper airway (UA) of patients with ALS through videofluoroscopy and to assess its applicability for a clinical trial.
Methods: In this cross-sectional study, participants with a diagnosis of ALS were enrolled to perform a single session of this RT. Epidemiological data and baseline assessment were collected. The outcomes assessed were kinematic variables from videofluoroscopy: retropalatal airspace size, narrowest airway, and pharyngeal area, during rest and RT.
Results: Eight participants were studied. During RT, an increase of 15% in the retropalatal airspace size was observed ($t = 5.14$, $p < 0.01$), along with a 123% increase in the narrowest airway ($t = -4.18$, $p < 0.001$) and 277% increase in the pharyngeal area ($t = -5.34$, $p < 0.001$).
Discussion: This is the first experimental study to show the effects of an RT on the UA using videofluoroscopy. RT had increased pharyngeal constriction, pharyngeal expansion, retropalatal airspace size, and the post-lingual narrowest airway during the intervention and proved to be of promise for a larger scale study. Considering the study results on acute UA constriction and expansion, the authors consider that this breathing exercise may have the potential to play a role in ventilation and as a treatment for pharyngeal-muscle weakness. A further clinical trial should be carried out to evaluate the chronic therapeutic effects of this technique and its impact on the clinical evolution of ALS.

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References

1. Plowman EK, Tabor-Gray L, Rosado KM, et al. Impact of expiratory strength training in amyotrophic lateral sclerosis: results of a randomized, sham-controlled trial. Muscle Nerve 2019;59(1):40–6.
2. Dorça A, Alcântara LA, Diniz DS, et al. Comparison between breath stacking technique associated with expiratory muscle training and breath stacking technique in amyotrophic lateral sclerosis patients: protocol for randomized single blind trial. Contemp Clin Trials Commun. 2020;19:100647.

CMS-11 Oral health status of ALS patients: a single-center observational study

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Introduction: In the disease scenario, most individuals with ALS have oral problems and difficulties in maintaining good oral health, either because of impaired orofacial function or weakness of the hands (1,2).
Objective: With little information and shortage of dentists in the multidisciplinary teams of ALS, the proposal is to assess the oral health conditions of patients with ALS, followed up in a reference center.
Methods: This single-center, cross-sectional observational study included 40 ALS patients, in Rio Grande do Norte, state in northeastern Brazil, between november 2020 and February 2021, evaluating their functional status using the ALSFRS-R and their oral health status through specific parameters, including an oral hygiene questionnaire and oral epidemiological examination (dental caries condition, periodontal and presence of change in soft tissue). The protocol was reviewed and approved by the Universidade Federal do Rio Grande do Norte Ethical Committee (The certificate of approval is CAAE 18628719.9.0000.5292.
Results: More than half of the patients brush their teeth less than 3 times a day and 75% do not floss. The mean DMF-d was 18 teeth (SD =7.29). Regarding periodontal conditions (CPI and PIP), the worst scores found were dental calculus (57.5%) and insertion loss was 0–3 mm (75%) and low percentage of periodontal pockets (no patient had pockets deep). There was no relationship between the clinical

phenotype and/or functional status of ALS with caries experience and periodontal conditions.
Discussion: The most important result of this study is the demonstration that most of them showed a poor oral status, which was independent from the functional status. In addition, the patients had absence of periodontal pockets, a marker of periodontitis. This data, not well understood, was also reported in previous studies (1,2). One intriguing finding is that pressure ulcers are either absent or only rarely occur in patients with ALS, even after such patients have spent long periods confined to bed and presenting severe motor limitations (3). The skin and gingival tissue derive from the same embryonic tissue. Thus, it is possible that ALS patients have a protective factor against the destruction of the skin and gum support tissues (4). Therefore, we concluded that all patients with ALS independently from the type of early stage symptoms needed assistance in performing oral hygiene measures.

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References

1. Nakayamaa R, Nishiyamaa A, Matsuda C et al. Acta Odontol Scand. 2018;76(4):294–8.
2. Tay CM, Howe J, Borromeo GI. Aust Dent J. 2014;59(3):309–13.
3. Ono S. Amyotroph Lateral Scler Other Motor Neuron Disord. 2000;1(3):191–9.
4. Paré B, Gros-Louis F. Rev Neurosci. 2017;28(5):551–72.

CMS-12 ALS Hope: a patient-created online dashboard of pre-symptomatic ALS research studies to better connect potential trial participants and the scientific community

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Background: Through frequent meetings with the scientific research community, the Familial ALS Community Team (fALST) – part of the patient-led organization I AM ALS – discovered that asymptomatic ALS research studies often have difficulty connecting with prospective study participants, significantly prolonging a study's time to completion. This is because pre-symptomatic gene carriers are hard to reach since they typically do not begin interacting with the ALS medical community until the onset of ALS symptoms. The ALS patient community was concerned that partially filled or slow-to-fill research studies may delay scientific progress and lead to reduced investment in future ALS research due to lack of adequate data points, both of which could be an impediment to finding a cure. Per industry research, the challenge of securing participants primarily stems from identifying an at-risk population and resolving various logistical and ethical considerations (1).
Methods: The fALST interviewed numerous principal investigators as well as both active and former pre-symptomatic study participants to identify ways to better match supply and demand with the intent of accelerating the time to conclude pre-symptomatic trials. The preliminary findings concluded that trial participants (supply) were most concerned with ethical and logistical study data points – whether the study included genetic counseling, their gene status would be documented in their medical records, the trial-related travel costs would be covered, and the length and frequency of trial visits would be feasible for them. Principal investigators and research coordinators (demand) were seeking ways to reach a wider audience within the difficult-to-reach pre-symptomatic ALS community, while having better upfront screening for potential trial eligibility and interest.
Results: The result was the creation of ALS Hope, a browsable online dashboard that contains pre-symptomatic research studies and the ability to filter each based on key attributes such as whether genetic counseling is included, travel is reimbursed, and the location and length of study details. Study information is either provided directly by the research coordinator, pulled from clinicaltrials.gov, or obtained directly from the research study's primary website. The dashboard is reviewed by the scientific advisory panel at I AM ALS to ensure it is eligible to be recommended by the medical community to their patients. The dashboard has received positive feedback from both patient and medical communities and has potential to accelerate the conclusion of pre-symptomatic research studies with the objective of more quickly finding a cure for ALS.
Conclusion: The fALST concluded that creating a user-friendly online dashboard of pre-symptomatic trials that included key trial attributes could significantly assist in matching potential research participants with ongoing and future research studies.

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Reference

1. Benatar M, Wuu J. Presymptomatic studies in ALS Rationale, challenges, and approach. Neurology. 2012;79:1732–9.

CMS-13 Interventions targeting psychological well-being for MND carers: a systematic review

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Background: Negative psychological impacts of caregiving for persons with MND are well-established and include carer burden, depression and anxiety. In recent years there has

been increased recognition and attention on interventions to improve the psychological well-being (PWB) of MND carers.
Introduction: This systematic review included interventions designed to improve the PWB of MND carers using quantitative, qualitative or mixed-method studies. An integrative approach to PWB was employed, encompassing both hedonic (the degree to which people experience positive emotions of feelings and happiness) and eudaimonic (functioning and purposeful behaviour) perspectives of the concept of PWB. The objectives of the systematic review were to (1) summarise current research, (2) assess the quality of evidence, and (3) evaluate the effectiveness of interventions.
Methods: A Mixed Methods Systematic Review was conducted based on Joanna Briggs Institute methodology for quantitative, qualitative and mixed methods reviews and Preferred Reporting Items for Systematic Reviews and Meta-Analyses guidelines. Outcomes of interest were any related to the PWB (hedonic or eudaimonic) of carers.
Results: Thirteen manuscripts (all published from 2013, 62% from 2017 onwards) met the inclusion criteria, including 12 studies (six mixed-methods, four quantitative, two qualitative). Four studies were randomised controlled trials, seven were uncontrolled longitudinal studies with a single treatment group and a pre-post design, and one was an observational survey. The 12 studies were conducted in a limited range of countries ($n = 4$). Critical appraisal revealed a wide range of methodological weaknesses. Only four of the ten studies with a quantitative component achieved summary quality scores above 50% with a lack of controls, power, blinding and low completion rates prevalent. Five of the eight studies with a qualitative component only obtained summary quality scores between 20% and 60% with incongruity between the stated philosophical perspective and the research methodology, no statement of epistemological and theoretical position, and no data analysis commonplace. Due to the heterogeneity of interventions, outcomes and measurements, a narrative and convergent approach to data synthesis was employed. A minority of studies demonstrated some benefits to hedonic and eudaimonic aspects of PWB, however the interpretability of these data was limited by methodological problems.
Discussion: The review revealed a low number of studies designed to improve the PWB of MND carers with the majority only published in the last four years. Benefits to hedonic and eudaimonic aspects of PWB were demonstrated in only a small number of manuscripts, most studies suffered from substantial methodological problems. Consequently, the overall evidence-base for interventions designed to promote PWB benefits for MND carers is still low. Given the well-known and considerable negative PWB impacts of the MND caregiving role, high-quality and carefully designed studies are urgently needed to facilitate the further development and testing of interventions promoting the PWB of MND carers.

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CMS-14 Genetic counselling regarding diagnostic genetic testing for ALS and FTD: results of a modified Delphi consensus survey

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Background: Access to genetic counselling and diagnostic genetic testing is a fundamental right of people living with ALS/MND (1). There are currently no consistent, evidence-based genetic counselling approaches or guidelines for diagnostic genetic testing in ALS/MND or frontotemporal dementia (FTD).
Objective: To identify areas of consensus and conflict on the ideal components of genetic counselling for diagnostic genetic testing in ALS/MND/FTD amongst health professionals and consumer stakeholders to inform the development of future consensus guidelines.
Methods: We used purposive and snowball sampling to recruit experts in the area of genetic counselling and testing for ALS/MND/FTD, including health professionals (e.g. clinical geneticists, genetic counsellors, neurologists, psychologists) and consumer experts (e.g. patients, relatives and staff from ALS/MND/FTD support organisations). An online, modified, multi-round Delphi consensus survey was conducted using REDCap. Items in the first round were informed by two systematic literature reviews and qualitative interviews with patients and relatives who had experienced diagnostic genetic testing. Descriptive and content analysis informed the development of the subsequent round and the final results.
Results: Forty-six experts participated: 100% completed round one, 95.65% ($N = 44$) completed round two. After round one, conflicts regarding the wording, content and timing of various items were identified. Items were updated based on feedback and presented for consensus in round two. After round two, a consensus was reached (>80% agreement) on all items. Sixteen items considered important as part of the diagnostic genetic testing process will be presented. These items focus on all aspects of genetic counselling service delivery, including providing information, counselling, and support. The results highlight that genetic counselling discussions should be adapted to the client's needs, which may be informed by social and family circumstances, their cognitive capacity, other medical needs, the pre-test likelihood of pathogenic variant detection and the subsequent genetic testing results.
Discussion: The emergence of genotype-driven therapy trials and higher than expected cases with a heritable basis means that genetic testing is becoming routine for individuals with ALS/MND/FTD. This research provides vital evidence of the high level of consensus amongst professional and consumer experts about the level of information, counselling and support required to inform genetic counselling and testing of these clients. We hope that the items developed will guide practitioners on the process of genetic counselling and testing, provide a basis for future research and inform the development of future consensus guidelines.

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Reference

1. The International Alliance of ALS/MND Associations. Fundamental rights for people living with ALS MND April 2021; 2021. https://www.alsmndalliance.org/wp-content/

uploads/2021/05/Fundamental-rights-for-people-living-with-ALS-MND-April-2021-Modified-May-6-pg.pdf

CMS-15 Diagnostic utility of Gold Coast Criteria in amyotrophic lateral sclerosis

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Objective: The diagnosis of amyotrophic lateral sclerosis (ALS) remains problematic, with current diagnostic criteria (revised El Escorial [rEEC] and Awaji) being complex and prone to error. Consequently, the diagnostic utility of the recently proposed Gold Coast criteria was determined in ALS.
Methods: We retrospectively reviewed 506 patients (302 males, 204 females) to compare the diagnostic accuracy of the Gold Coast criteria to that of the Awaji and rEEC criteria (defined by the proportion of patients categorized as definite, probable, or possible ALS) in accordance with standards of reporting of diagnostic accuracy criteria.
Results: The sensitivity of Gold Coast criteria (92%, 95% confidence interval [CI] = 88.7–94.6%) was comparable to that of Awaji (90.3%, 95% CI =86.69–93.2%) and rEEC (88.6, 95% CI =84.8–91.7%) criteria. Additionally, the Gold Coast criteria sensitivity was maintained across different subgroups, defined by site of onset, disease duration, and functional disability. In atypical ALS phenotypes, the Gold Coast criteria exhibited greater sensitivity and specificity.
Interpretation: The present study established the diagnostic utility of the Gold Coast criteria in ALS, with benefits evident in bulbar and limb onset disease patients, as well as atypical phenotypes. The Gold Coast criteria should be considered in clinical practice and therapeutic trials.

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CMS-16 Trends in communication board use by amyotrophic lateral sclerosis patients in Japan

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Background: A transparent communication board with several letters has been used as a unique communication method for Japanese amyotrophic lateral sclerosis (ALS) patients. The communication board is accessible to patients with preserved eye movement. However, it requires skill from both the patient and caregiver.
Objectives: This survey aimed to understand the trends in the use of communication boards for ALS patients through the crude tabulation of a web-based questionnaire survey among primary caregivers.
Methods: A total of 1977 home nursing stations across Japan with at least seven nurses and more than 50 patients were asked to participate in the survey. Home-visiting nurses provided primary caregivers of ALS patients with a QR code that directed to a web-based questionnaire, which included the patient's age, sex, ALS Functional Rating Scale (ALSFRS-R), communication board usage, reasons for using the communication board, and with whom they used it. The Osaka University Clinical Research Review Committee approved the survey (#20394).
Results: From 7 May to 19 July 2021, 96 responses were obtained. Excluding eight duplicated responses, 88 responses were analysed. There were 49/88 (56.8%) patients receiving invasive tracheostomy ventilation and 17/88 (19.3%) receiving non-invasive positive pressure ventilation. The median ALSFRS-R score was 10.5 (interquartile range: 6 to 17.25). The median time from diagnosis was six years (interquartile range: 3 to 9 years). Communication boards were used by 45 (51.1%) patients, but not by 43 (48.9%). Among those using communication boards, 12 (26.7%) patients were introduced to them in hospitals, and 27 (60%), in the community (by home-visiting nurses, healthcare assistants, and public-health nurses). Communication boards were used by 31 (68.9%) patients to communicate with family members, including spouses and children, while 40 (88.9%) used them to communicate with nurses, and 28 (62.2%), with healthcare assistants.
Discussion: More than half of the ALS patients used communication boards. As the results were obtained from patients receiving care from home-visiting nurses, we received limited responses from patients in the early stages of diagnosis and those who independently performed activities of daily living. The survey findings suggested that the communication board was a significant, augmentative, and alternative communication modality for Japanese patients, especially home-care patients with advanced ALS symptoms. Furthermore, community-based healthcare professionals, such as home-visiting nurses and healthcare assistants, played a significant role in introducing and using communication boards.

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CMS-17 A new measure of disease severity of amyotrophic lateral sclerosis by conversion of forced vital capacity

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Background: The evaluation of disease severity in ALS patients requires two perspectives: the degree of functional decline and the rate of progression. Forced vital capacity (FVC) is a major indicator representing the functional state of

respiratory muscle. The decline pattern of FVC is not uniform and not necessarily linear during the ALS course (1,2), and thus it is impossible to compare the rate of disease progression by FVC value itself. It is reported that FVC at a first measurement was a significant predicator of survival and disease progression (3), suggesting that the rate of disease progression can be evaluated from FVC value by consideration of the duration from onset to measurement.
Objective: The purpose of this study is to develop a new measure representing the rate of disease progression by conversion of FVC to scores corrected for duration from onset to measurement, named duration-adjusted FVC.
Methods: Development of the duration-adjusted FVC was performed using longitudinal FVC data of the Pooled Resource Open-Access ALS Clinical Trials (PRO-ACT) Database (6522 measurements from 939 patients). We performed a nonparametric approach to generate the duration-adjusted FVC so that patients had consistent scores throughout ALS progression. Association between the duration-adjusted FVC and ALS prognostic factors was examined using the PRO-ACT database and our single-centre cohort data (96 patients). Furthermore, correlation between the duration-adjusted FVC and the Risk Profile proposed by the Treatment Research Initiative to Cure ALS (TRICALS) which was based on the well-validated survival model of European Network to Cure ALS (4) was examined.
Results: FVC was converted to the duration-adjusted FVC in the range of 35–106 depending on FVC and duration from onset to measurement. Low duration-adjusted FVC was associated with bulbar onset, older age at onset, and short survival, even when patients of equivalent FVC were compared. Furthermore, the duration-adjusted FVC was significantly correlated with the Risk Profile of TRICALS ($R^2 = 0.90$, $p < 0.001$)
Discussion: Low duration-adjusted FVC was associated with factors that indicate fast progression and high duration-adjusted FVC was associated with factors that indicate slow progression. The duration-adjusted FVC, determined from a single FVC, will be a useful tool to assess the rate of disease progression in individual patients.

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References

1. Ackrivo J, Hansen-Flaschen J, Jones BL, et al. Am J Respir Crit Care Med. 2019;200(12):1513–21.
2. Sato K, Iwata A, Kurihara M, et al. J Neurol Sci. 2019;403:7–12.
3. Jahandideh S, Taylor AA, Beaulieu D, et al. Amyotroph Lateral Scler Frontotemporal Degener. 2018;19:294–302.
4. Westeneng H-J, Debray TPA, Visser AE, et al. Lancet Neurol. 2018;17(5):423–33.

CMS-18 neuroprotective hormone levels in ALS: a cross-sectional study

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Background and objective: Various hormones of the HPA axis have been studied for their role in ALS. However, each of these studies have evaluated only a few hormones. The effects of hormones are interdependent and influence each other directly or indirectly. So, our objective was to study multiple hormones in ALS. Testosterone, Estradiol (E), Progesterone (P), Dehydroepiandrosterone Sulphate (DHEAS), Growth Hormone (GH) provide neuroprotection (1–3). Stress being a factor, Cortisol levels need to be studied.
Method: 23 patients with probable/definite ALS. Plasma Total Testosterone (TT), Estradiol, Progesterone, DHEAS, GH, and Cortisol were tested at 8am by Chemiluminescence. Severity was assessed on the revised ALS Functional Rating Scale (ALSFRS-R) and King's staging. Z-score was calculated for TT and DHEAS using age-matched reference values. For other hormones laboratory assay range was used to compute z scores. Percentage analysis was performed. Spearman's Rank Correlation was performed on DHEAS levels vs ALS FRS R score.
Results: Of 23 patients there were 18 males, 5 females; 17 limb onset, 6 bulbar onset; 19 King's Stage 3. Percentage analysis showed TT levels(66.7%< mean;44.4%< 1 + SD);E(44.4%< mean; 22.2%< 1 + SD); P(100%< mean); DHEAS (95.7%< mean;43.5%< 1 + SD); GH(94.4%< mean;83.3%< 1 + SD);Cortisol(94.4%< mean;66.7%< 1 + SD). We found a statistically significant, moderately positive correlation between DHEAS z-score and ALS FRS R($r = 0.49$, $p = 0.018$).
Discussion and conclusion: All the neuroprotective hormones studied were lower than normal levels in the majority of ALS patients.DHEAS levels declined with disease progression. Since, majority patients were in King's Stage 3, low Cortisol levels may indicate adrenal exhaustion. This study indicates that replacement of a single hormone may be insufficient and multiple hormones need to be considered while planning an effective therapeutic strategy.

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References

1. Sane H, Varghese R, Paranjape A, et al. Correlation of testosterone levels with progression of amyotrophic lateral sclerosis in males: a cross-sectional study. Adv Res J Multidiscip Discoveries. 2019;37(4),22–7.
2. Quinn TA, Robinson SR, Walker D. Dehydroepiandrosterone (DHEA) and DHEA sulfate: roles in brain function and disease. Sex Horm Neurodegener Proc Dise. 2018;3:41–68.
3. Chung JY, Sunwoo JS, Kim MW, Kim M. The neuroprotective effects of human growth hormone as a potential treatment for amyotrophic lateral sclerosis. Neural regeneration research. 2015;10(8):1201.

CMS-19 Study of the visual pathway with diffusion tensor imaging and cognition in patients with amyotrophic lateral sclerosis

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Background: Retinal involvement and changes in visual evoked potentials have been detected in ALS patients, suggesting that the visual pathway from the eyes to the brain may be involved in ALS. However, there is a lack of imaging studies examining the visual pathway in ALS.
Purpose: We aimed to explore the microscopic changes of visual pathway in ALS patients using DTI, and to detect the changes in peripapillary retinal nerve fiber layer (RNFL) thickness using OCT. Moreover, we analyzed the relationship between the DTI measures of visual pathway and cognitive status, disease duration, disease severity and RNFL thickness in ALS patients.
Methods: The study included 33 ALS patients and 20 healthy controls from August 2019 to January 2021. We used DTI to measure FA and ADC values of visual pathway, including the optic nerve, optic chiasm, optic track, lateral geniculate body and optic radiation. The RNFL thickness was measured with OCT. The disease severity was evaluated by ALSFRS-R. The cognitive status was evaluated via ECAS. We compared the DTI measures of visual pathway and RNFL thickness between ALS-ci (ALS patients with cognitive impairment) group, ALS-nci (ALS patients without cognitive impairment) group and healthy control group. Pearson or Spearman correlation analysis was performed to explore the correlation between DTI measures and RNFL thickness, disease duration, and ALSFRS-R score in ALS patients.
Results: Compared with healthy controls, FA values of the whole visual pathway were significantly increased and ADC values of the optic radiation were significantly decreased in ALS group, and the RNFL was thicker in ALS group. We speculated that this may be a compensatory neural connection enhancement of the visual pathway in ALS patients. The increase in FA values of some parts of the visual pathway in ALS-ci were not as remarkable as that in ALS-nci, and the increase of FA values were more remarkable in ALS patients with normal visuospatial ability than ALS patients with visuospatial impairment. This suggested that ALS patients with cognitive impairment, especially visuospatial impairment, have reduced visual pathway compensatory capacity. In ALS patients, FA values of optic chiasma were positively correlated with RNFL thickness, and ADC values of optic chiasma and lateral geniculate body were negatively correlated with RNFL thickness; FA values of optic nerve were negatively correlated with disease duration; ADC values of optic radiation were negatively correlated with ALSFRS-R.
Conclusion: Our study indicated that compensatory changes may occur in retina and visual pathway of ALS, and the DTI changes of visual pathway of ALS patients were correlated with cognitive function, disease duration and severity of disease.

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CMS-20 Information of patients from ongoing post-marketing surveillance, evaluating the real-world safety and effectiveness of edaravone for amyotrophic lateral sclerosis patients in Japan (SUNRISE Japan)

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Background: Edaravone, a free-radical scavenger, was developed and approved for the treatment of patients with amyotrophic lateral sclerosis (ALS) in various countries from 2015 to 2021 (Japan, June 2015; South Korea, December 2015; United States, May 2017; Canada, October 2018; Switzerland, January 2019; China, July 2019; Indonesia, July 2020; Thailand, April 2021). The approvals were based on efficacy and safety data from patients with a definite or probable ALS diagnosis (1). Edaravone demonstrated statistically significant efficacy in slowing the progression of ALS, as assessed by scores on the ALS Functional Rating Scale-Revised (ALSFRS-R). However, to date, other end points, such as survival time or time to tracheal intubation, have not been assessed. Therefore, this ongoing study, SUNRISE Japan, aims to obtain real-world data about the long-term effectiveness, including survival endpoints, and safety of edaravone in ALS patients in Japan.
Objectives: To assess the safety and effectiveness of edaravone through post-marketing surveillance for patients with ALS.
Methods: Overall, more than 800 ALS patients who were edaravone treatment – naïve have been enrolled in the surveillance program and will be followed up for 5 years. Safety assessments include adverse events up to 1 year. Efficacy assessments include duration of survival and duration until invasive tracheal intubation is needed up to 5 years; clinical events such as introduction of tube feeding, gastrostomy, and intermittent noninvasive ventilator assistance up to 1.5 years; and ALSFRS-R scores up to 1.5 years. The survey is being conducted in accordance with Japan's Ministerial Ordinance on Good Post-marketing Study Practice.

Results: Seven hundred ninety-nine patients were included in both the safety and efficacy analysis sets. Adverse drug reactions occurred in 97 patients (12.14%), and serious adverse drug reactions occurred in 30 patients (3.75%). The most common serious adverse drug reaction was "Hepatic function abnormal" which occurred in 6 subjects (0.75%). The changes in ALSFRS-R score were 38.5 ± 6.6 ($N = 726$) at baseline, 32.6 ± 10.0 ($N = 444$) at 6 months later and 29.7 ± 11.1 ($N = 320$) at 12 months later, respectively. In this report, no unexpected safety signals were seen, nor any inconsistencies with the clinical trials.
Discussion: This program may be useful in understanding longer-term effects of edaravone in the treatment of ALS in Japan and across the globe, as well as serving as a model for other long-term surveillance studies in neurodegenerative diseases.

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Reference

1. Writing Group on behalf of the Edaravone (MCI-186) ALS 19 Study Group. Lancet Neurol. 2017;16:505–12.

CMS-21 Predictors of the need for non-invasive ventilation (NIV) during gastrostomy insertion in patients living with motor neurone disease (MND)

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Introduction: Motor neuron disease (MND) is a multi-system disease that affects the brain, nerves, and muscles, including those of the respiratory system, with progressive weakness and risk of respiratory failure (1).
The incidence of long-term gastrostomy in patients with MND is 20–40% (2), and those with respiratory muscle involvement need close monitoring during gastrostomy insertion.
Objectives: To assess whether various respiratory parameters can predict the need for non-invasive ventilation (NIV) cover during Percutaneous Endoscopic Gastrostomy (PEG)/ Radiologically Inserted Gastrostomy (RIG) insertion.
Method: We retrospectively analysed the electronic records of 172 patients undergoing PEG/RIG insertion at the University Hospitals of North Midlands (UHNM) between 2016 and 2019. The patients were admitted electively for PEG/RIG insertion with NIV cover following outpatient assessment by the respiratory team. A respiratory nurse specialist accompanied patients to the endoscopy suite and applied NIV in the event of significant oxygen desaturation or drowsiness. Statistical analysis was conducted using SPSS.
Results: Of the 172 patients: 93 patients (54%) were men, and the mean age was 67.6 years. A total of 157 patients had a prior diagnosis of MND, with 152 (97%) of those having successful PEG or RIG insertion. 93% had PEG insertion, while 7% had RIG insertion. The mean length of hospital stay was 8.83 days. A total of 105 patients required NIV cover during PEG insertion. Of the cohort on domiciliary NIV, 89.4% required NIV cover compared to 50% of those MND patients not on long-term NIV. 67% of patients with $PaCO_2 < 6$ needed NIV support compared to 87% of those with $PaCO_2 >= 6$. Patients with $PO_2 < 10$ were more likely to require NIV (73.7%) compared to those with $PO_2 >= 10$ (59%).
Conclusion: Patients living with MND and already established on NIV, those with $PaCO_2 >= 6$, and patients with $PO_2 < 10$ were more likely to require NIV cover during PEG/RIG insertion. Thus, this study supports the need for close monitoring for respiratory symptoms, regular lung function tests & blood gases, and to involve the respiratory team before PEG/RIG insertion, especially in the presence of respiratory symptoms.

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References

1. Rafiq M, Proctor AR, Mcdermott CJ, et al. Respiratory management of motor neurone disease: a review of current practice and new developments. Pract Neurol. 2012;12:166–76.
2. Sarkar P, Cole A, Scolding NJ, et al. Percutaneous endoscopic gastrostomy tube insertion in neurodegenerative disease: a retrospective study and literature review. Clin Enosc. 2017; 50(3):270–8.

CMS-22 Respiratory interventions in a population-based ALS cohort: demographics and survival determinants

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Background. The gold-standard for respiratory failure treatment in ALS is represented by non-invasive mechanical

ventilation (NIMV) and eventually, in the latest phases of the disease, by invasive mechanical ventilation via tracheostomy (IMV).
Aims. To assess the outcome of ventilatory support in a large population-based cohort, evaluating the characteristics of patients who chose either or both NIMV and IMV and to determine their prognostic determinant.
Methods. The study population includes ALS patients identified through the prospective population-based register Piemonte and Valle d'Aosta Register for ALS (PARALS). For the present study we considered ALS cases diagnosed from 2008 to 2015 ($N = 1159$). In order to assess the variations over time in the use of respiratory support, we compared the data of the 2008–2015 period to the preceding 8 years (2000–2007) ($n = 1070$).
Results. A total of 375 (32.4%) patients underwent NIMV alone, 82 (7.1%) performed NIMV and then underwent IMV (NIMV + IMV), 82 (7.1%) underwent IMV without previous NIMV, and 620 (53.5%) did not receive any ventilatory support. The frequency of patient who received a respiratory support significantly increased from 37.9% to 46.5% compared to the 2000–2007 period ($p = 0.0001$).
A. The median survival time after NIMV initiation to either IMV or death was 1.00 year (IQR 0.49–2.33), significantly increased compared to the previous period. FVC% resulted to be the strongest determinant of NIMV outcome, followed by age, and lower ALSFRS-R mean monthly decline, and higher ALSFRS-R upper limb subscore, all considered at time of NIMV.
B. The median survival time after IMV initiation to death or the censoring date was 1.94 (IQR 0.64–5.05) with an increase compared to the 2000–2007 period (1.23, IQR 0.36–3.89; $p = 0.01$). According to Cox multivariable analysis, factors related to a better survival after IMV initiation were younger age at time of IMV, lower ALSFRS-R% decline before IMV, to be married, and previous use of NIMV.
Discussion. In this large population-based study, we have found that the frequency of use of NIMV and IMV increased over time, highlighting the improved adherence to the standard guidelines for the treatment of respiratory impairment in ALS Among the factors related to the outcome after respiratory mechanical support, the most relevant were patients' age, ALSFRS-R decline before the intervention, and FVC%. These real-world data will be useful for designing clinical trials for novel therapies in ALS, which should keep into account the substantial effect of mechanical ventilation on the course of the disease and its demographic and clinical determinants.

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CMS-23 Delayed diagnosis and diagnostic pathway of ALS patients in Portugal: where can we improve?

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Introduction: Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disease with unsatisfactory treatment options. Early diagnosis is important in order to provide patients best management and opportunity for enrolment in clinical trials. However, ALS diagnosis can be challenging, with resultant significant diagnostic delay. Analysis of the diagnostic pathway and identification of the causes of diagnostic delay is imperative.
Methods: A cohort of 580 ALS patients followed in our ALS clinic in Lisbon was studied. Demographic, disease and sociocultural factors were collected. Time from first symptom onset to diagnosis, time from first symptoms to first consultation, specialist's assessment and investigation requested were analyzed. Predictors of diagnostic delay were evaluated by multivariate linear regression, adjusting for potential confounders.
Results: We included 580 ALS patients (mean age of 65 ± 12 years). Most patients were classified as probable ALS according to the revised El Escorial criteria; 22% of patients were diagnosed with progressive muscular atrophy. One fifth of patients had a bulbar onset. The median diagnostic delay from first symptom onset was 10 months (IQR =5–18 months). Bulbar onset and faster disease progression were associated with a lower probability of diagnostic delay (bulbar onset: coef. −10.54, $p < 0.001$; fast progression: coef. −6.50, $p < 0.001$, respectively). Lower annual income was associated with longer diagnostic delay (coef. 5.25, $p = 0.003$). The majority of patients were first assessed by non-Neurologists (80%), namely General Practitioners. The median time from first medical observation to diagnosis was 6 months (IQR =2–11). The majority (70%) of patients had 2 or 3 medical evaluations before diagnosis was achieved, and almost 15% consulted 4 or more specialists. Patients who were evaluated by a Neurologist had an increased likelihood of being correctly diagnosed, decreasing time to diagnosis. Almost all specialists who made the diagnosis (95%) requested the Electromyography (EMG).
Conclusions: Late referral from non-Neurologists to a Neurologist is a potentially modifiable factor contributing to significant diagnostic delay. Educational interventions targeted to non-Neurologists physicians, in order to increase awareness of ALS and, consequently promote early referral to a Neurologist at a tertiary center, will be important in reducing diagnostic delay.

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CMS-24 User-centred design and testing of a bespoke online toolkit (www.NIV4MND.co.uk) for healthcare professionals learning about non-invasive ventilation in motor neuron disease

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Background: Non-invasive ventilation (NIV) improves the quality of life and survival in MND but many patients fail to receive effective ventilation and service provision varies across the UK. Based upon previous research and expert focus groups, an evidence-based educational toolkit (www.niv4mnd.co.uk) has been developed to educate healthcare professionals (HCPs) about NIV in MND care.
Objectives: To further develop and optimise NIV4MND in line with user-centred design principles.
Methods: A mixed-methods approach was employed. Data was collected from HCPs across the UK via a questionnaire ($n = 16$) and semi-structured 45-min interviews ($n = 7$). Questions in the survey centred around design, content, usability, and future use. Topics covered in interviews followed similar themes but were iteratively expanded based upon results of the survey and previous interviews. Quantitative data produced were analysed using SPSS descriptive statistics and qualitative data underwent thematic analysis using NVivo. The study was approved by the University of Sheffield Research Ethics Committee.
Results: Thematic analysis showed very positive overall views on the website. Participants reported that it represented available evidence and agreed best practice. They felt it filled a gap in the current education landscape and that they would recommend the resource to colleagues as well as use it themselves in the future. Key areas for improvement were noted as reducing large walls of text, increasing visibility of good practice guidelines, future maintenance of the toolkit, increasing video accessibility, and improving accessibility on mobile devices. These changes will be implemented into the site.
Discussion: The website clearly reflects the key messages that experts feel are important with these results allowing us to validate the content of the toolkit. Within the field it will act as a complementary site to www.mybreathing.co.uk, which provides similar information for patients and carers of those considering NIV. The site was felt to most useful for those who are inexperienced in providing NIV in MND care to allow them to better understand the roles of different members of the team and the full pathway of care. The results indicate that HCPs believe this toolkit will improve the quality of care given to patients which should hopefully lead to the ultimate goal of better survival and quality of life for people with MND.

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CMS-25 Self-administered ALSFRS-R on the telehealth in MND (TiM) system: initial evaluation of validity in Irish users

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Background: The ALSFRS-R is a widely used 12-item ordinal scale, incorporating key constructs relevant to the progression of disease in ALS/MND. The recent shift to remote monitoring highlighted the requirement for valid and reliable patient-reported outcome measures. Previous studies have shown validity of a self-administered ALSFRS-R versus standard ALSFRS-R (1). The self-administered ALSFRS-R on the Telehealth in MND (TiM) system was adapted from the ALSFRS-R using patient-friendly wording (2). The measure requires evaluation of validity compared with standard clinician administered ALSFRS-R.
Objectives: To analyse the validity of the self-administered, TiM ALSFRS-R compared with clinician-administered ALSFRS-R.
Methods: Patients included in the study attended the MND multidisciplinary clinic in Beaumont Hospital, Dublin and used the TiM telehealth system. TiM ALSFRS-R responses, completed monthly via telehealth, were compared with clinician-administered ALSFRS-R responses. Corresponding scales completed within one month of each other were included. Agreement between TiM and clinic ALSFRS-R scores were examined using Bland-Altman plots and correlation analysis.
Results: Twenty-eight patients completed 114 TiM ALSFRS-Rs between January and June 2021. Twenty-eight pairs of scores from both TiM and clinician-administered ALSFRS-r for 22 patients were included in the analysis. Average time between

self-administered and clinic ALSFRS-R was 6 days (range 0–30 days). The mean difference in total ALSFRS-R score was 0.86 (SD3.5) points, 0.79 points (SD1.6) in the bulbar subscore, -0.04 points (SD1.32) in the upper limb subscore, -0.11 points (SD1.03) in the lower limb subscore and -0.14 points (SD2.05) in the respiratory subscore points. Particular disagreement was noted in the dyspnoea question in patients using Non-Invasive Ventilation (NIV). The total TiM and clinic administered ALSFRS-R scores showed high correlation and internal consistency (Spearman's rho =0.82; Cronbach's alpha 0.93).
Discussion: Our findings indicate good agreement between self-administered and clinician-administered ALSFRS-R total scores. Use of NIV incurs an automatic score of 0 on the dyspnoea question according to the ENCALS ALSFRS-R SOP, which resulted in disagreement as patients responded to the question based on symptoms. Based on these findings as well as clinician and patient feedback on wording, the TiM ALSFRS-R has been further modified and future evaluation of scale validity will include a larger sample size.

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References

1. Manera U, Cabras S, Daviddi, M, et al. Validation of the Italian version of self-administered ALSFRS-R scale. Amyotroph Lateral Scler Frontotemporal Degener. 2021;22(1–2):151–53.
2. Hobson EV, Baird WO, Bradburn M, et al. Using telehealth in motor neuron disease to increase access to specialist multidisciplinary care: a UK-based pilot and feasibility study. BMJ Open. 2019;9(10):e028525.

CMS-26 "It's a big nugget of information that I don't know what to do with": information and support needs of people living at an increased genetic risk of MND

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Background and objectives: In light of increasing understanding on the genetic basis of MND, and clinical trials targeting particular genetic variants, there have been discussions around the benefits of (routine) genetic testing for people with MND. This could see more relatives living with the knowledge that they are at an increased genetic risk of developing MND in the future, in spite of family history. Living with the knowledge of genetic risk can have a profound impact on people's lives. This study focuses on the information and support needs of these individuals.
Methods: This study draws on interviews with 36 people affected by inherited forms of MND (including 7 people with MND, 4 partners, and 25 people at an increased risk of developing symptoms in the future, the principal focus of this study). Interviews were analysed thematically, using a method of constant comparison. Interviews were carried out as part of PhD research on family experiences of inherited MND.
Results: Living at an increased genetic risk of MND raises unique concerns and challenges, and as such, people may need help in making sense of and managing this information and the decisions it raises. This study explores critical junctures where people had a particular need for information and support, including around diagnostic and pre-symptomatic genetic testing, and significant life events in the family such as the death of relatives, or sharing information on genetic risk with children. It outlines forms of support that might address some of these needs, including emotional and informational support, and ongoing monitoring.
Discussion: This study aims to highlight the profound yet diverse impact that living at an increased genetic risk of MND can have on people's lives, and the information and support that would be helpful, which varies markedly between individuals and over time. This is not a caution against genetic testing of people with MND but rather an attempt to better understand the information and support needs of family members.

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CMS-27 Evaluation of the nation-wide implementation of ALS Home Monitoring & Coaching: an e-health innovation for personalized care for patients with ALS

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Background: With the e-health innovation 'ALS Home monitoring & Coaching' (ALS H&C) patients with ALS can be monitored remotely. ALS H&C was successfully implemented in UMC Utrecht in 2017 and has recently been implemented in 10 other multidisciplinary ALS care teams in the Netherlands.
Objectives: The aim of this study was to evaluate the implementation success/failure and – process and usability of the

e-health innovation according to patients and healthcare professionals.
Methods: We implemented ALS H&C in 10 ALS care teams in the Netherlands using a participatory action research approach with a strong foundation in implementation science theory (1). Three months after implementation, implementation success was evaluated as well as its determinants. Therefore, several implementation outcomes were evaluated based on fieldnotes and an online survey (evaluation framework by Proctor and telemedicine technology acceptance model of Chau) that was administered to healthcare providers and patients, including satisfaction, usability, sustainability (whether the teams continued using ALS H&C after three months use) and fidelity (whether the innovation and implementation plan was executed as intended).
Results: Patients ($n = 71$) and healthcare providers ($n = 76$) were satisfied with the care concept (median 8.0 and 7.0, resp., on a scale from 1 to 10). Patients were positive about the usability, but healthcare providers, especially those with little experience with the platform, considered the usability to be below-average. In three teams, the implementation was not sustainable. The reported main reason for stopping was related to usability (platform not integrated with their electronic health record system) and it appeared that there were fidelity issues in these teams (e.g. the healthcare professional who would be working with ALS H&C the most did not attend all preparatory meetings with the project group, they did not adhere to the healthcare protocol for the monitoring, meetings were not well attended by the healthcare team).
Discussion: The present study demonstrates that ALS H&C can be successfully implemented in multidisciplinary ALS care using a participatory action research approach with a strong foundation in implementation science theory. However, it was not successful in every team. Results showed that usability and fidelity played an important role in implementation failure. Therefore, to improve implementation success, healthcare providers should receive sufficient training on how to use the innovation and it is recommended to integrate the e-health innovation with the local electronic health record systems. In addition, although participating teams are allowed to slightly adjust the innovation and implementation plan to the local context, core elements of the innovation and implementation plan need to be included.

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Reference

1. Dontje ML, Kruitwagen-van Reenen E, Visser-Meily JMA, et al. Implementation and evaluation of an e-health innovation for personalized care for patients with amyotrophic lateral sclerosis (ALS): protocol for a participatory action research study. Implement Sci Commun. 2021;2(1):25.

CMS-28 Exploring the inclusivity of telehealth for people living with motor neuron disease, and the validity of a telehealth version of the ALSFRS-R

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Background: Access to specialist MND services improves the outcomes of people with MND (PwMND), however physical and geographical barriers hinder access to these services. The Telehealth in MND (TiM) system is an online digital care system allowing PwMND and healthcare providers to track symptoms via questionnaires such as the ALS functional rating scale revised (ALSFRS-R). With the recent increase in use of telehealth in clinical practice, it is important to ensure new service configurations are inclusive. Here we explore potential barriers to an inclusive implementation of telehealth. The ALSFRS-R has high inter and intra-rater reliability, and has been previously validated for self-administration. This study aims to validate the self-administered ALSFRS-R on TiM (ALSFRS-R-T).
Methods: Sociodemographic factors, health status, and MND specific factors – as outlined in the NIHR INCLUDE framework – were assessed against uptake and adherence to the TiM system. Adherence was calculated comparing individual registration time on TiM, to the number of ALSFRS-R-T completions – a 100% adherence is one ALSFRS-R-T completion every 14 days. A multi-centre validation of the ALSFRS-R-T was performed in two MND care centres (Sheffield, UK and Dublin, Ireland). PwMND who recently completed an ALSFRS-R-T were contacted to take part in an ALSFRS-R phone interview. Scores from the phone interviews were compared against the PwMND's most recent ALSFRS-R-T completion using Pearson's correlation and a Bland-Altman plot.
Results: Full results from 2020 to 2021 will be presented. Currently 93 PwMND were invited to use TiM and 59 PwMND are enrolled. Age, sex, gastrotomy and site of MND symptom onset did not influence uptake of telehealth. NIV use was more common in those that enrolled on TiM compared to those that did not enrol ($X^2(1, N = 59) = 4.57$, $p = 0.033$). Age, sex, gastrostomy, ventilation usage, rate of MND progression, site of MND symptom onset and King's staging were not associated with adherence levels. However, there was a positive correlation between ALSFRS-R scores and adherence ($\rho = 0.477$, $p < 0.001$). Initial validation of the ALSFRS-R-T demonstrates high correlation between TiM and phone ALSFRS-R ($\rho = 0.980$, $p < 0.001$). A Bland-Altman plot suggests no presence of systemic directional bias. Calculated King's staging using ALSFRS-R-T showed high reliability, with an intraclass-coefficient of 1.00 compared to King's staging calculated from phone interviews.

Discussion: Uptake of PwMND to a telehealth system was not affected by age or gender. We did see a higher uptake in those using NIV, which may be due to the additional challenges those with respiratory failure experience in getting to clinic or the increasing complexities of their care needs. Future research will explore how specific functional impairments affect uptake and use of telehealth systems.

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CMS-29 Interpreting the meaning of existence for the person with motor neurone disease and their family carer(s)

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The genesis of this thesis lies in experiences of caring professionally for people living and dying with motor neurone disease (MND) and in personal experiences of caring for my mother with the same condition. These experiences led me to an understanding that MND threatens the sense of existence for both the person with the condition and their family. MND is a devastating neurodegenerative terminal condition that results in loss of the motor neurones that enable a person to move, speak, and at the end of life, to breathe. At the outset of this project, previous research had focused on the narratives of people living with MND at a single point in time; thus knowledge of the meaning of existence with MND through time is limited.

This is a multiphase study that uses two distinct methodologies: hermeneutic (interpretive) phenomenology and autoethnography to explore the lived experiences of self and others. The three key areas of study are (i) the phenomenon of existence when someone is given a diagnosis of MND and in the context of receiving healthcare; (ii) the meaning of living with uncertainty for people diagnosed with MND; and (iii) the meaning of supporting a loved one with MND as they die. The outcomes of this research have significantly contributed to knowledge through the publication of six peer-reviewed papers. The findings of the research are collated and integrated to develop a person-centred model of care that emphasises the need for MND professionals to acknowledge the temporal aspects of caring for a person with MND, and for their family carers.

The thesis concludes that MND care is complex, but that this complexity can be reduced if the range of professionals who provide MND care interpret the care needs of the people at end of life with MND to provide not only multi-professional but inter-professional person-centred care.

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CMS-30 The complexity of planning care at a multidisciplinary motor neurone disease (MND) clinic

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Background: Regular review at expert multidisciplinary clinics (MDCs) is advocated for people with MND/ALS (1) and a survival benefit has been demonstrated (2). Access to expert clinicians at an MDC, working collaboratively during one visit is a significant advantage, particularly for those travelling long distances. However, this model of care can result in long visits.

Objectives: To better understand the objective patient experience at the MDC, through documentation of the patient pathway through the clinic, the demand upon services and ability to plan appointments in advance, with an ultimate goal of development of solutions to maximise efficiency.

Methods: This study was conducted at the national MND MDC in Beaumont Hospital, Dublin between November 2020 and July 2021. Firstly, time-motion observational studies of a random sample of patients attending the clinic were carried out, to document the clinic journey. Secondly, patients identified in advance as requiring consultation with specific HCPs ('flagged') were recorded and compared with the HCP consultations that actually occurred.

Results: During this period there were 289 in-person clinic attendances and 291 remote consultations. All patients had a consultation with a doctor and clinical nurse specialist, who determined the need for referral to other clinic HCPs. Fifteen time-motion studies of patients attending in person were recorded. On average, patients spent a total of 172 min in clinic, 97 min waiting, and 75 min in consultations (45% of visit). Of the 289 in-person attendances, patients saw between 1 and 5 HCPs per visit. Thirty-two percent of occupational therapist ($n = 79$), 42% of speech and language therapist ($n = 74$), 96% of dietitian ($n = 49$) and 29% of physiotherapist ($n = 116$) consultations were flagged in advance.

Discussion: The complex care needs of MND patients, which change rapidly, is a significant challenge in the provision of MDT MDC care. This study illustrates that the ability to predict or schedule consultation needs in advance of clinic is limited and demonstrates the requirement for flexibility in care provision. Remote monitoring and telehealth could assist in more effective planning, reducing waiting time for patients and their families, and maximising clinicians' use of time. Due to the busy nature of the MDC, all clinician interactions may not have been captured.

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Acknowledgements

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References

1. NICE. Motor neurone disease: assessment and management. In: Excellence NIFHAC, ed. Clinical guideline NG42. London: NICE; 2016.
2. Rooney J, Byrne S, Heverin M, et al. A multidisciplinary clinic approach improves survival in ALS: a comparative study of ALS in Ireland and Northern Ireland. J Neurol Neurosurg Psychiatry. 2015;86:496–501.

CMS-31 Impact of COVID-19 on the functional status of ALS patients: a Chart Review

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Background: In patients with ALS, follow-up in clinical care centers improves quality of life (1) including mental quality of life (2). The international COVID-19 pandemic caused restrictions of in-person healthcare services in order to prevent further spread of the virus. This and subsequent lockdowns precluded most in-person multidisciplinary clinic visits. The pandemic guidelines also caused cessation of valuable therapies including physical, occupational, speech and respiratory therapy. Besides a greater vulnerability in case of infection, ALS patients are also more vulnerable to the indirect effects of a quarantine as they are highly dependent on others (e.g. caregivers, respiratory therapists, dieticians, rehab therapists and medical providers) to manage their disease progression and maintain the best possible health status (3).
Objective: To study the extent to which clinical care changes due COVID-19 lockdown and social distancing impacted overall self-perceived functioning of ALS patients seen in a NYC ALS clinic and survey whether certain demographic and/or disease-specific features are underlying causes of functional decline.
Methods: A retrospective chart review from 1 January 2020 to present day of 50 PALS between the ages of 18–90 at Hospital for Special Surgery is being completed. Medical records of these PALS are being reviewed to assess for change in functionality during COVID-19 lockdown. Revised ALS Functional Rating Scale (ALSFRS-R) assessment were performed both during in-person and telemedicine clinic visit appointments. Significance of change is defined as 6+ points on the ALSFRS-R scale during COVID-19 lockdown.
Results: Preliminary results suggest a strong correlation between lack of home services (home health aides(HHA), home rehabilitation therapies), outpatient therapeutic services and functional decline. A preliminary trend indicates that patients who reported the most functional changes over the quarantine period were the same patients who had to halt all home services (HHA and home rehabilitation therapy).
Conclusion: Our findings support recent studies showing COVID-19-related challenges compromised ALS clinical care and research (4).

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References

1. Traynor BJ, Alexander M, Corr B, et al. Effect of a multidisciplinary amyotrophic lateral sclerosis (ALS) clinic on ALS survival: a population-based study, 1996–2000. J Neurol Neurosurg Psychiatry. 2003;74:1258–61.
2. Van den Berg JP, Kalmijn S, Lindeman E, et al. 2005.
3. Cabona C, Ferraro PM, Meo G, et al. Predictors of self-perceived health worsening over COVID-19 emergency in ALS. Neurol Sci. 2021;42:1231–36.
4. Andrews JA, Berry JD, Baloh RH, et al. Amyotrophic lateral sclerosis care and research in the United States during the COVID-19 pandemic: challenges and opportunities. Muscle Nerve. 2020;62(2):1.

CMS-32 "More than a 'patient advocacy' organization": patient-centred perspectives of non-profit ALS/MND health charities

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Background: Non-profit ALS/MND health charities (HCs), such as the provincial and national ALS societies in Canada, provide important support to people with ALS (PwALS) and family caregivers. While the roles of HCs in disease advocacy and research funding are well represented in the literature (1), patient-centred investigation of how PwALS and caregivers perceive the ALS/MND societies has not been explored.
Objective: To identify how ALS/MND HCs are perceived by and support PwALS and family caregivers.
Methods: Drawing on data from the ALS Talk Project, an asynchronous, moderated focus group study using the online itracksTM platform, we investigated the ways in which the Canadian ALS societies influenced the experiences of PwALS and caregivers. PwALS and caregivers participated in separate focus groups. Moderated discussion took place over 14 weeks. Topics included: the diagnostic journey, living/coping with ALS, information needs, research participation, and advanced care planning. We also asked questions about participants' experiences with the ALS societies as appropriate and analyzed all statements/discussion related to the societies. Qualitative data was exported in NVivo 10TM and inductively analyzed using directed content analysis and the constant-comparative approach.
Results: Fifty-seven PwALS and 43 caregivers from five Canadian provinces participated in seven focus groups. We identified three primary themes. (1) The medical clinics and ALS societies were overwhelmingly perceived as counterparts in providing care ("the clinic and ALS society have played a huge part in helping us"), with the ALS societies playing an important role as healthcare navigators ("We had trouble navigating homecare. The ALS society was helpful and now we have a homecare caseworker"). The ALS society also filled needs not addressed by the clinics ("the ALS Society referred me to a phycologist to help me deal with this diagnosis"). (2) Participants identified specific supports provided by the societies, including peer support groups, equipment loans, disease information, and personal understanding ("much

needed emotional support and reassurance that I was not alone"). (3) Participants understood that the societies had a role in research, but primarily identified their roles facilitating understanding of research ("great for dissemination of the latest research outcomes") and fundraising ("funding and research are obviously really important").
Discussion: Non-profit ALS/MND HCs play a pivotal role by providing client services and support to PwALS, as well as critically assisting PwALS/caregivers navigate within the medical system. Canadian PwALS and caregivers view the ALS societies as integral to the provision of medical care and rely on the ALS societies for information and knowledge translation.

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Reference

1. Johnston WS, Luth W. Public policy of ALS in Canada. In: Blank RH, Kurent JE, Oliver D, eds. Public policy in ALS/MND care. New York: Palgrave; 2021:77–96.

CMS-33 Measuring sleep and well being in caregivers: data from a study of children and youth carers in ALS/MND

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Background: Children and youth under the age of 19 "young carers", provide daily care for family members living with illness (1). Prior research shows caregiving affects school performance, social support, stress, and anxiety across populations (2), and specifically in ALS families (3). Yet, little information exists detailing how care may disrupt sleep, leading to other potential negative health effects, critical to overall caregiver well-being.
Objectives: (1) Establish feasibility of recruitment, retention, and compliance in young carers and a non-caregiving comparison group.
(2) Assess differences in sleep and well-being variation and patterns between young carers and non-caregiving controls.
Methods: Quasi-experimental matched comparison study used a sample of age and gender matched young caregivers ($n = 8$) and non-caregivers ($n = 12$), to assess the feasibility of measuring sleep quality in young carers, and identify initial differences in sleep quality and patterns between young caregivers and non-caregivers. Participants completed a pre/post survey including the Pittsburgh Sleep Quality Index (PSQI), wore an actigraphy device (GeneActive), and journaled sleep/wake times for 5 consecutive 24 h periods.
Results: Both groups completed the surveys and wore the actigraphy devices 99.2% of the time, over a 5-day period. Data from the PSQI revealed shorter sleep duration ($t = 51.19$ (11.99)), efficiency ($t = 55.49$ (14.00)), poorer overall sleep quality ($t = 51.32$ (12.26)), and higher rates of utilizing medications to help them sleep ($t = 50.81$ (11.49)) in caregivers. Objective sleep data using the GENEActiv device reflected the subjective data finding caregivers had lower total sleep time (CG $= 6.75 \pm 1.47$, NCG $= 7.08 \pm 1.36$) and had lower sleep efficiency than non-caregivers ($.80 \pm .23$). Case examples of various sleep patterns are reported across groups.
Discussion: Study results demonstrate feasibility while providing crucial initial data on sleep quality in young carers. Findings underscore the need to better document the impact of caregiving on young carer well-being across areas, including sleep. Implications exist for larger scale studies examining how sleep disruption impacts well-being broadly, as well as measuring the impact of care in the movement, informing support and respite interventions for young carers across disorders.

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Acknowledgements

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References

1. Hunt G, Levine C, Naiditch L. Young caregivers in the U.S.: findings from a national survey. Bethesda, MD: National Alliance for Caregiving [in collaboration with United Hospital Fund]; 2005.
2. Leu A, Becker S. A cross-national and comparative classification of in-country awareness and policy responses to 'young carers'. J Youth Stud. 2017;20(6):750–62.
3. Kavanaugh MS, Cho CC, Howard M, et al. US data on children and youth caregivers in amyotrophic lateral sclerosis. Neurology. 2020;94(14):e1452–59.

CMS-34 Measuring self-reported fatigue in people with ALS

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Background: Patient reported outcome measures are of increasing importance for ALS research. Fatigue is a prevalent, bothersome, and undertreated symptom in patients with amyotrophic lateral sclerosis (ALS). Measuring fatigue is challenging because multiple factors may contribute to the patient experience of fatigue, such as tiredness, mental fatigue, and motor fatigue. A number of self-reported fatigue scales exist and may capture fatigue in people with ALS (PALS) in different ways.
Objective: The data collected in this study will be used to (1) inform future studies quantifying self-reported fatigue in ALS and (2) correlate self-reported fatigue to self-reported mood and function in ALS.
Methods: PALS consented and enrolled themselves online. They were asked to complete self-reported questionnaires at baseline, 4-weeks, 3-, 6-, 9- and 12-months. The study is

ongoing. We report results of the baseline through 6-month follow-up. The following surveys were included: Revised ALS Functional Rating Scale (ALSFRS-R), Rasch Overall ALS Disability Scale (ROADS), Neurological Fatigue Index - Motor Neuron Disease (NFI-MND), Fatigue Severity Scale (FSS), ALS Depression Inventory (ADI-12), and Epworth Sleepiness Scale (ESS). Pearson correlations were used to compare baseline fatigue and functional scores. 6-month changes in fatigue and functional status were also evaluated.
Results: Currently, 182 PALS completed baseline surveys, and 120 have completed the 6-month survey. At baseline, both the ALSFRS-R and ROADS demonstrated a significant negative correlation with the NFI-MND ($r = -0.27$ and -0.28, respectively) and the FSS ($r = -0.25$ and -0.24, respectively). The NFI-MND demonstrated significant positive correlations with ADI-12 and ESS ($r = 0.38$ and 0.28, respectively). At 6 months, the per month change in ALSFRS-R continued to demonstrate a significant negative correlation with NFI-MND ($r = -0.27$) however the ROADS did not show a significant correlation. Full results will be presented.
Discussion/Conclusions: Initial findings suggest that weak correlations exist between self-reported measures of fatigue and function. There was also a weak correlation between measures of fatigue and both depression and sleepiness. These initial findings illustrate the complexity of measuring fatigue. They suggest that fatigue is impacted by more than the single factors of function, depression, and sleepiness, and warrants further evaluation.

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CMS-35 Impact of occupational therapy services delivered via telehealth to patients with amyotrophic lateral sclerosis

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Objective: To identify the impact of occupational therapy services delivered via telehealth to patients with amyotrophic lateral sclerosis (ALS) and recommend future use of telehealth in this patient population.
Introduction: Telehealth is an emerging practice area for occupational therapy practitioners being used across practice areas. There is little evidence surrounding use of telehealth with patients with ALS by occupational therapists.
Method: Semi-structured interviews were conducted with six participants about their experience with occupational therapy delivered through telehealth. Interviews were recorded and transcribed, with a thematic analysis performed afterwards.
Results: The following four themes were identified: valued accessibility, activity analysis insight, engagement, and tiers of occupational therapy, with 12 inter-related subcategories. Participants perceived it as a positive that telehealth allowed convenient access to OT services allowing them control and flexibility over their care. Telehealth was not perceived positively as an initial visit for OT or during periods of change in functional status.
Conclusions: This study revealed occupational therapy practitioners should use telehealth as a means to deliver occupational therapy services to patients with ALS in conjunction with face to face visits as a hybrid. An initial face to face visit is recommended to obtain baseline measures and introduce occupational therapy to this population to build rapport. This study identified an underlying hierarchy of occupational therapy services with face to face visit being perceived as 'top tier', and telehealth being perceived as a secondary delivery of service. Occupational therapists should consider utilizing telehealth in patients with ALS when their functional status has plateaued to a stable and predictable state. Telehealth can be used to monitor patients and provide means for passive learning. If there is a change in status, physical or cognitive, occupational therapists should initiate a face to face visit over a telehealth visit. A face to face visit is more beneficial for parallel learning and to acutely evaluate a patients functional status through physical testing. Further research should investigate the use of occupation during intervention delivered via telehealth with this population.

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CMS-36 Anthropometry and body composition for nutritional and prognostic evaluation of patients with amyotrophic lateral sclerosis

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Background: Malnutrition is prevalent in patients with Amyotrophic lateral sclerosis (ALS) and negatively affects patients' quality of life and survival. Nutritional assessment and monitoring are key components of the nutrition care process.
Objectives: To compare nutritional assessment parameters between patients and controls and to investigate possible associations between these parameters in ALS patients.
Methods: Cross-sectional study carried out with ALS patients treated at the Neuromuscular Disease Care Program in Natal, Brazil. The functional status was assessed by the ALS Functional Rating Scale-Revised (ALSFRS-R). Phase angle and body composition data were obtained through bioimpedance analysis and software (Quantum II®, RJL Systems). Energy intake was evaluated through the application of two 24-h food records. Among all the parameters, fat mass index (FMI), fat-free mass index (FFMI), percentages of fat mass (FM) and fat-free mass (FFM), bone mineral content (BMC), skeletal muscle mass (SMM) were of primary interest in this analysis. Also, daily energy intake (DEI) and daily energy deficit (DED) were considered. The GraphPad Prism 6 software was used for the statistical analysis of the data. A p-value ≤ 0.05 was considered significant. The study was approved by the HUOL Ethics Committee (CAAE 40467214.0.0000.5292).
Results: Thirty-eight participants with a mean age of 53.1 ± 12.2 were included in this study (21 ALS patients and 17 healthy controls). Most of the patients had spinal ALS as an initial clinical manifestation (67%) and ALSFRS-R score ≤ 24 points (62%). Most of the parameters were significantly different between patients and controls, including PA. The DEI was significantly lower among patients compared to controls, but the DED was not significantly different, since the basal metabolic rate and the daily energy expenditure were lower in ALS patients. Regarding the associations among the parameters used, we observed moderate to strong correlations between PA and ALSFRS-R, and SMM and PA, among

others. The BMC had a strong negative correlation with BMI and FMI.
Discussion: The clinical features and physiopathology of ALS explain the impairment of the nutritional status of these patients compared to the control group. The association between PA and functional status corroborates its use as a prognostic indicator in ALS patients. Among the parameters investigated, PA, SMM, BMC, and FMI stand out due to the number and intensity of significant correlations found. These parameters can be useful to monitor the prognosis and the nutritional status at the time of diagnosis and during follow-up.

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References

1. Barone M, Viggiani MT, Introna A, et al. Nutritional prognostic factors for survival in amyotrophic lateral sclerosis patients undergone percutaneous endoscopic gastrostomy placement. Amyotroph Lateral Scler Frontotemporal Degener. 2019; 20(7–8):490–6.
2. Roubeau V, Blasco H, Maillot F, et al. Nutritional assessment of amyotrophic lateral sclerosis in routine practice: value of weighing and bioelectrical impedance analysis. Muscle Nerve. 2015;51(4):479–84.

CMS-37 A feasible and safe alternative: radiologically inserted gastrostomy with limited sedation for people with amyotrophic lateral sclerosis

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Background: People with ALS (PALS) with a forced vital capacity (FVC) $< 50\%$ or abnormal overnight pulse oximetry have higher risk of respiratory failure when undergoing procedures requiring respiratory depressant anesthetics, including gastrostomy tube placement (1). Radiologically-inserted gastrostomy (RIG) tubes with local anesthesia and limited sedation offer an alternative for PALS, including those with advanced disease who may not tolerate endoscopy.
Objective: Determine the feasibility and outcomes of RIG tube placement with limited sedation in PALS.
Methods: Subjects in this single-center registry had a 16 Fr or 18 Fr RIG tube placed without intubation or endoscopy, followed by overnight observation. Procedural anesthesia was determined by the anesthesia team. Demographic, clinical, procedural, and outcomes data were retrospectively collected for at least 90 days. Descriptive statistics were summarized as percentages for categorical variables and means with ranges for continuous variables. A Spearman correlation was calculated for the relationship between ALSFRS-R score and death within 90 days post-procedure.
Results: Twenty-eight PALS had a mean age of 65 years (46–84) at time of RIG placement. Sixteen (57.1%) were female. Most common disease presentation was bulbar (53.5%). Average FVC was 46% of predicted (23%-73.26%) and 15 (53.6%) individuals had abnormal pre-procedural overnight oximetry reported as sleep disordered breathing (25.0%), gas exchange abnormality (10.7%), or both (17.9%) preceding RIG. Four used noninvasive nocturnal ventilation. Average time between ALS symptom onset and RIG tube placement was 673 days (249–1922). Mean pre-procedural ALSFRS-R was 29.4 (7–43). Lidocaine and bupivacaine were used as local anesthetics. Thirteen received dexmedetomidine hydrochloride (DH) alone; 13 received DH plus fentanyl, propofol, midazolam, and/or ketamine; 2 received fentanyl and propofol with or without midazolam. Tube placement failed in 1 (3.5%), leaving 27 for post-procedural outcomes analysis. Mean pre-procedural pain score was 0.3 (0–6); 4-h post-procedural was 0.8 (0–4); and at hospital discharge was 0.9 (0–8). Thirty-day readmission rate for non-procedure-related issues was 25.9%. Six subjects had a 30-day minor complication: 1 (3.7%) tube dislodgement, 1 (3.7%) peri-tube leakage, and 4 (14.8%) superficial peristomal infection. No PALS had a 30-day major procedure-related complication. Thirty-day and 90-day mortality was 5 (18.5%) and 10 (37.0%), respectively, and were not procedure-related, including hyperglycemia in 2 (20%) and presumed respiratory failure in 8 (80%) including 2 after transition to hospice. The mean pre-procedural ALSFRS-R in individuals who died within 90 days was lower than the mean of those alive beyond 90 days [24.8 versus 31.5, p-value 0.04, $r = -0.33$].
Conclusions: RIG placement with local anesthesia and limited sedation is a feasible and safe option for PALS, including those with more advanced disease. Future investigation will focus on the feasibility and safety of a same-day procedure.

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Reference

1. Bote SM, Martinez NP, Amarilla CE, et al. Respir Care. 2020; 65(8):1128–34.

CMS-38 Remote cough monitoring for predicting bulbar and respiratory impairment

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Background: Early identification and monitoring of the progression of bulbar and respiratory impairment is critical to providing quality care for patients diagnosed with ALS (PALS). For many patients, regular clinic visits are challenging. As such, remote monitoring for healthcare practitioners to objectively monitor health status is needed. Cough impairment is commonly reported by PALS, is a promising feature for tracking function and has been linked to respiratory, speech, and swallowing impairments. Our long-term goal is to determine the utility of remote cough monitoring for the

identification and monitoring of bulbar and respiratory impairment in PALS.
Objective: To assess the accuracy of a machine learning analysis of volitional cough recordings collected remotely to classify PALS with or without bulbar and respiratory impairment.
Methods: Cough data were collected from 69 PALS and 6 healthy controls using the Beiwe smartphone research platform. Participants completed the Revised ALS Functional Rating Scale (ALSFRS-R) and recorded a volitional cough on the Beiwe app. Using machine learning (ML), participants were classified into binary categories (symptomatic or asymptomatic) based on ALSFRS-R bulbar and respiratory subscale scores (12 or <12), as well as on the speech and swallowing questions (4 or <4). Acoustic features from a single voluntary cough from each participant were extracted using the OpenSMILE ComParE13 configuration. Highly correlated features were removed, and feature selection was performed within each cross-validation testing set, retaining only those features that differed between the groups of interest at alpha =0.05 level. Logistic regression models were used for classification. Results were cross-validated using the leave-one-out approach.
Results: Based on ALSFRS-R scores, 49 PALS presented with bulbar and/or respiratory impairment, 42 presented with bulbar impairment (mean score =5.2, range =1–10), 32 presented with respiratory impairment (mean score =6.1, range =0–11), 34 presented with speech impairment (mean score =1.9, range =0–3), and 27 presented with swallowing impairment (mean score =1.4, range =0–3). ML algorithms detected: (1) bulbar or respiratory impairment with an area under the curve (AUC) = 0.72, (2) bulbar impairment using with AUC =0.79, and (3) speech impairment with AUC =0.82. The algorithm did not accurately classify by swallow (AUC =0.47) or respiratory function (AUC =0.45).
Discussion: Preliminary findings indicate that with voluntary cough recordings, ML algorithms are able to detect differences between PALS with and without bulbar impairments with high accuracy. Interestingly, this finding appears to be driven by ALSFRS-R speech impairment scores, perhaps because the ALSFRS-R may be less responsive to respiratory and swallowing change. Further investigation is warranted to (1) determine whether volitional cough may serve biomarker of bulbar and respiratory impairment onset and progression, and (2) validate this work with perceptual ratings from clinicians.

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CMS-39 ALS telehealth in a multidisciplinary service in Natal, Brazil

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Background: The multidisciplinary following was compromised during the COVID-19 pandemic. Telehealth had already been use for many neurologic areas showing good results. During this time, it emerged as an urgent need to maintain the health care during social distance.
Objective: To evaluate the feasibility of telehealth for the provision of multidisciplinary ALS care. Also, assess the acceptability of telehealth among patients and caregivers.
Methods: Retrospective cohort study. Multidisciplinary evaluations were perform using the Teleconsulta platform during the period of April 2020 to March 2021, at Hospital UniversitarioOnofre Lopes, Natal, Brazil. The patients included had ALS and at least one in person clinical evaluation. The patients and the caregivers answered satisfaction questionnaires.
Results: There were 46 patients, 32 male and 14 female. 78% of patients had ALSFS <39 and 22% was >39. The average distance from the residence to the reference services was 120 km. It was 72 consultations. The mean time attendance duration was 63 min. The evaluation was positive to very positive on the majority of questions.
Discussion: The telehealth is viable and positive in terms of satisfaction. It was even more positive for patients with advanced disease or livingin a city far from the referral center. It can be a form of follow-up to alternate with in person consultations or even replace standard care for cases with advanced disease.

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References

1. Haulman A, Geronimo A, Chahwala A, Simmons Z. The use of telehealth to enhance care in ALS and other neuromuscular disorders. Muscle Nerve. 2020;61:682–91.
2. Capozzo R, Zoccolella S, Musio M, et al. Telemedicine is a useful tool to deliver care to patients with amyotrophic lateral sclerosis during COVID-19 pandemic: results from southern Italy. Amyotroph Lateral Scler Frontotemporal Degener. 2020; 21(7–8):542–48.
3. Andrews JA, Berry JD, Baloh RH, et al. Amyotrophic lateral sclerosis care and research in the United States during the COVID-19 pandemic: challenges and opportunities. MuscleNerve. 2020;62(2):182–86.
4. Van De Rijn M, Paganoni S, Levine-Weinberg M, et al. Experience with telemedicine in a multidisciplinary ALS clinic. Amyotroph Lateral Scler Frontotemporal Degener. 2018; 19(1–2):143–48.

CMS-40 Genetic testing experiences in a tertiary ALS center

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Background: The availability of commercial genetic testing in the United States has increased over the past decade with the discovery of new genes associated with ALS. Targeted genetic treatments are increasingly available in clinical trials and genotype-phenotype presentations of ALS are being

identified. As a result, genetic testing for both familial and sporadic ALS has potential clinical implications and prognostic value for pALS and family members (1).
Objectives: The goal of the current study was to determine the incidence of ALS genetic variants in a clinic-based population.
Methods: 121 individuals with ALS were offered genetic testing during their routine multidisciplinary care visit. All patients were offered C9orf72 repeat expansion testing followed by reflex testing with a multigene panel. Descriptive statistics were used to summarize results by family history (fALS, sALS) and incidence of genetic variants.
Results: Genetic testing was completed in 97% (117/121) of individuals with ALS who were offered testing. Of those that completed testing, 35% ($n = 41$) yielded positive results for genes associated with ALS (2). The incidence of pathogenic ALS variants was 12% with the majority being C9orf72 ($n = 10$) followed by SOD1 ($n = 4$). Of those with C9 and SOD1 positive results, 90% (9/10) and 100% were familial ALS, respectively. The incidence of variants of uncertain significance (VUS) was 66% ($n = 27$) and included: SETX ($n = 7$), SPG11 ($n = 7$), OPTN ($n = 4$), SQSTM1 (n =3), GRN ($n = 2$), CHMP2B ($n = 2$), MAPT ($n = 1$) and TBK1 ($n = 1$). Second risk variants included: C9orf72/SETX ($n = 1$), OPTN/SPG11 ($n = 1$) and CHCHD10/GRN ($n = 1$).
Discussion/Conclusions: ALS genetic testing was accessible to the majority of our ALS clinic patients. Although there are limited consensus and management guidelines for genetic testing in ALS, our clinic has found value in genetic testing for ALS disease management and prognosis, particularly for eligibility in targeted treatment trials (3).

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References

1. Wagner KN, Nagaraja HN, Allain DC, et al. Mol Genet Genomic Med. 2018;6(2):224–9.
2. Mejzini R, Flynn LL, Pitout IL, et al. ALS genetics, mechanisms, and therapeutics: where are we now? Front Neurosci. 2019;13:1310.
3. Roggenbuck J, Fong JC. Clin Lab Med. 2020;40(3):271–87.

CMS-41 The Italian version of the Rasch-Built Overall Amyotrophic Lateral Sclerosis Disability Scale (ROADS): a validation study

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Background: The ALS Functional Rating Scale-Revised (ALSFRS-R) is the most widely used tool for the clinical monitoring in ALS patients. Despite his usefulness as a multidimensional scale, the combined score derived from different domains is not linearly related to symptoms severity. The Rasch-Built Overall ALS Disability Scale (ROADS) has recently been developed and validated in different languages (1,2) to overcome these limitations.
Objectives: To validate the Italian version of the ROADS scale and assess the reliability of its administration to patients versus their respective caregivers and the correlation to the corresponding ALSFRS-R.
Methods: In the Turin ALS Centre, the ROADS Scale questionnaire was administered together with ALSFRS-R to 55 ALS patients during regular follow-up assessments. The same questionnaire was administered also to the caregivers instructing them to compile it according to their own assessment of the patients' functional status. Correlation analysis was performed using Spearman's rho, Bland-Altman difference plots, Cronbach's alpha coefficient and Intraclass correlation coefficient (ICC), one-way random effects were used for proper comparison.
Results: The median normalized ROADS values did not differ significantly between patients and caregivers (73.0, IQR 60.0–86.0 vs 70.0, IQR 57.0–82.0, $p = 0.524$). Their correlation coefficient was found to be very high (ICC 0.95, $p < 0.001$; Cronbach's alpha coefficient 0.94) and the total score agreement based on Bland-Altman showed no systematic directional bias (mean difference: 0.82, 95% limits of agreement: −15.9–17.6). Stratifying for age, sex, site of onset, type of caregiver, disease duration, and progression rate, we found high ICC values that did not change significantly among the considered categories. We also found a high correlation between ROADS and ALSFRS-R total score (patients' correlation coefficient: 0.88; caregivers' correlation coefficient: 0.87).
Discussion: We validated the Italian version of the ROADS scale, confirming its reliability when administered either to patients or their caregivers. We confirmed also that ROADS showed high reliability also when administered to caregivers; in addition, stratification for different patients and caregivers features, such age, sex, site of onset, type of caregiver, disease duration, and progression rate, did not alter significantly the concordance metrics.
Conclusions: The Italian version of the ROADS scale is a valid and reliable tool to monitor disease burden, showing a high level of agreement between the responses given by patients and caregivers.

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References

1. Fournier CN, Bedlack R, Quinn C, et al. JAMA Neurol. 2020;77(4):480–8.
2. Sun C, Fournier CN, Ye S, et al. Chinese validation of the Rasch-Built Overall Amyotrophic Lateral Sclerosis Disability Scale. Eur J Neurol. 2021;28(6):1876–83.

CMS-42 A multicentre evaluation of saliva management in people with MND (ProSec3 study)

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Background: Problems caused by oral secretions are common in people with MND. They represent a significant burden for patients with little evidence to guide management.
Objectives:
1. To describe how common saliva problems are in the UK MND population
2. To describe how saliva problems are currently treated
3. To identify the effectiveness of each treatment for saliva problems
4. To identify which, and how often, drugs for saliva problems cause side effects and lead to discontinuation
Methods: The study was open to all adults diagnosed with ALS/MND with the capacity to give consent. Saliva, drug and medical history data were collected alongside four measures (amended clinical saliva score for MND (CSS-MND (1)), ALSFRS-R, a global change questionnaire and modified Likert scale) at 1–5 visits held at approximately 3-month intervals.
Results: ProSec3 was a prospective, cross-sectional cohort study with a longitudinal component. 504 participants were recruited from 36 sites (February 2018–September 2020). Based on ALSFRS-R visit 1 data, 60% of participants (271/455) (95% CI: 55%, 64%) reported saliva symptoms. This proportion increased over the visits, with 75% (54/72) (95% CI: 64%, 84%) reporting saliva problems at visit 5. We also estimated the incidence of thick and thin saliva problems. At visit 1, 24% (108/456) (95% CI: 20%,28%) reported thin saliva problems, 9% (41/456) (95% CI: 7%,12%) reported thick and 25% reported both thick and thin saliva issues (115/456) (95% CI: 22%, 30%). Anticholinergics were by far the most commonly prescribed drugs for saliva management ($n = 261$), followed by mucolytics ($n = 86$), with other therapies such as moisteners, botulinum toxin and cannabinoids reported in small numbers. Saliva medications were reported as discontinued 164 times. Inadequate symptom control was cited as a reason for discontinuation of therapy 57 times and a range of side effects were frequently cited. The most commonly reported side effects related to gastrointestinal issues ($n = 83$), leading to the discontinuation of medication 76 times. The high discontinuation rate of these drugs indicates poor effectiveness, which will be explored further when the longitudinal data are analysed prior to the symposium.
Discussion: This is a large survey investigating the management and prevalence of saliva problems in people with MND. We have identified that the drugs commonly being prescribed to alleviate secretion problems are often not effective and many have unpleasant side effects. As such, the study provides empirical data confirming a need to investigate better methods of managing saliva problems in this patient group.

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Reference

1. McGeachan A, Hobson E, Shaw P, et al. Amyotroph Lateral Scler Front Degener. 2015;16(1–2):108–13.

CMS-43 Prospective evaluation of respiratory chest infections in a European ALS cohort: results from the REVEALS Study

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Background: Respiratory function is an important predictor of survival in ALS and is measured regularly, informing decisions on interventions such as ventilatory assistance (NIV and cough augmentation techniques). Few prospective longitudinal studies exist that focus on respiratory tract infections and their clinical correlates. The Registry of Endpoints in ALS (REVEALS) study was a prospective longitudinal study in 6 European sites which had a secondary aim of prospectively examining RTI rates and morbidity. This abstract presents findings on RTIs and their clinical correlates.
Methods: This was a prospective observational study which took place from January 2018 to February 2021. Participants with baseline Kings Stage 2 or 3 ALS were assessed in clinic at three-monthly intervals. Respiratory measures included SNIP, FVC, SVC and PCF. Data relating to respiratory history, respiratory symptoms was also collected. Between clinic visits, presence of chest infections was prospectively ascertained by text, mail or phone every two weeks. Where a chest infection was reported, the patient was interviewed about diagnosis, hospitalisation, antibiotic use and symptomatic burden.
Results: 280 (66.8% male, Mean age 63.3, SD11.6) participants with baseline Kings Stage 2 (59.6%) or 3 (40.4%) were recruited to the study. Time since diagnosis was 7.05 ± 17.45 months, onset was Bulbar in 19% and Spinal in 81%. In total, participants responded to 5013 chest infection checks (mean 15: range 1–57 per participant) over 55 ± 38 weeks. A mean of 1.69 RTIs (range 1–7) were reported by 78 (27.8%) participants. Diagnosis was made by a GP/Hospital in 69%, and hospitalisation required in 14%. Antibiotics were prescribed in 55%. Difficulty clearing secretions was reported in 83% with over a fifth reporting significant difficulty and 8% an inability to clear secretions. Infections had a duration of 9.6 ± 7.5 days. At baseline, there were no significant differences in PCF, %predicted SVC, FVC or SNIP between those who developed a chest infection during the study and those that did not ($p > 0.05$).
Discussion: The incidence of respiratory tract infections measured prospectively was high at 27.8%. Over 1 in 10 required hospitalisation and significant difficulty was reported with clearing secretions, suggesting a need for respiratory therapy during RTIs.
Conclusion: Respiratory infections are a significant cause of morbidity in ALS. Routinely performed baseline respiratory measures did not predict risk of infection.

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Reference

1. Tattersall R, Murray D, Heverin M, et al. Respiratory measurements and airway clearance device prescription over one year in amyotrophic lateral sclerosis. Amyotroph Lateral Scler Frontotemporal Degener. 2020;21(1–2):70–7.

CMS-44 Exploring the impact of the COVID-19 pandemic on caring for people living with amyotrophic lateral sclerosis

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Background: The immediate impact of the COVID-19 pandemic on amyotrophic lateral sclerosis (ALS) (otherwise known as motor neuron disease) care was that services providing testing, non-invasive ventilation (NIV) and gastrostomy were negatively affected (1). There is a need to explore the experiences of people living with ALS (plwALS) and healthcare professionals (HCPs) caring for plwALS during the pandemic to see how the provision of care has changed and the impact of this.
Objective: To identify how ALS care has been affected by the global pandemic from the perspectives of plwALS and HCPs (both at an individual and care centre level).
Methods: Three separate surveys were carried out to explore the experiences of plwALS, HCPs and ALS care centres during the pandemic. Closed-ended questions were analysed using descriptive and inferential statistics. Free text responses were analysed thematically. The data were triangulated to provide an in-depth look at the impact of COVID-19 on ALS care.
Results: Fifty-three plwALS, 74 HCPs and 23 care centres completed the surveys. Five main themes were identified: negative emotions, keeping safe, losses, delivering care and alternative care delivery in a pandemic. PlwALS reported feeling isolated and worried about contracting COVID-19 and the majority opted to stay at home ($n = 46$, 88%). Changes to care included longer waiting times for appointments and face-to-face appointments being cancelled or replaced by telephone or video consultations. Whilst benefits of this were reported (e.g. keeping safe and removing the need for travel), plwALS and HCPs raised concerns about not having face-to-face contact (e.g. communication difficulties, incomplete clinical assessments and HCPs missing symptoms). Provision of testing such as respiratory function testing, treatments such as riluzole and interventions such as NIV and gastrostomy were also delayed/disrupted. Moreover, plwALS had less access to multidisciplinary services such as physiotherapy, nutrition and speech and language therapy. PlwALS and HCPs reported that patients were not always getting adequate care because of changes to ALS care.
Discussion and conclusions: PlwALS and HCPs reported that care had been disrupted which resulted in patients receiving sub-optimal care. Some changes that were reported are not in line with published clinical guidance (e.g. delays in testing and delays in receiving treatments) (2). We have developed several recommendations. These include adopting new service delivery models, offering home visits, and using telehealth. Our recommendations are likely to be helpful for ensuring continuity of care both in a pandemic and a non-pandemic context and both nationally and internationally.

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References

1. Glasmacher SA, Larraz J, Mehta AR, et al. J Neurol. 2020.
2. National Institute for Health and Care Excellence. (2016) Motor Neurone Disease: Assessment and Management. NICE guideline (NG42).

CMS-45 Multimodal dialog based speech and facial biomarkers capture differential disease progression rates for ALS remote patient monitoring

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Background: Continuous monitoring of ALS symptoms is crucial to improve quality of life because both survival and progression rate can vary substantially between individuals. Speech and facial biomarkers can serve as useful proxies for disease progression and can be monitored remotely and automatically.

Objective: The main objective of this longitudinal study is to analyze acoustic and facial speech metrics extracted from a web-based conversational assessment over time. Our hypothesis is that such multimodal remote patient monitoring allows us to measure and track changes in certain speech and facial biomarkers (i) more frequently and cost-effectively, while (ii) remaining as informative (if not more) than current clinical standard scales in capturing differences in ALS disease progression between slow and fast progressors.
Methods: Speech and video data were collected from 56 people with ALS (pALS) using a cloud-based multimodal dialog platform between October 2020 and July 2021. In addition to structured speech tasks such as sentence repetition and diadochokinesis (DDK) tasks, participants also filled out the ALSFRS-R, a standard scale for monitoring ALS progression. Based on these ALSFRS-R scores, we stratified participants into two cohorts: (a) slow progressors (<0.47 points/month decline; 18 males, 19 females), and (b) medium to fast progressors (≥0.47 points/month; 12 males, 7 females). The rate of change was calculated based on the first and last observation for each participant. Acoustic (timing, frequency, and energy-related) and visual features (movement, surface, and velocity-related) were automatically extracted from the recordings. Their rate of change was computed as linear regression slopes over time. To normalize for sex-specific differences in metrics (such as fundamental frequency), we z-scored all metrics by sex group. Statistical tests were conducted to identify features for which these slopes are significantly different between the two cohorts.
Results: Acoustic speech features related to timing (percent pause time), frequency (fundamental frequency), and voice quality showed statistically significant differences in their rate of change between slow and fast progressors at a significance level of 0.05. Effect sizes (Cohen's *d*) were moderate for most features. Among facial kinematic features, we observed significant differences for the change over time in higher order statistics of the jaw and lips (such as acceleration of the jaw center). Effect sizes were moderate. Additionally, we show that splitting up the analysis by different speech task types (such as read speech, DDK, or vowel phonation) yields a higher responsiveness of certain metrics as measured by effect size.
Conclusions: Our findings demonstrate the efficacy of remote patient monitoring via a cost-effective and scalable dialog platform to extract informative speech and facial biomarkers, allowing the potential capture of information that standard scales like the ALSFRS-R (which is not very granular) might not register.

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CMS-46 Is dysphagia a predictive factor for survival in amyotrophic lateral sclerosis patients with severe respiratory failure?

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Background: Amyotrophic lateral sclerosis (ALS) is a rapidly progressive motor neuron disorder that causes respiratory failure and dysphagia (1). Percutaneous endoscopic gastrostomy (PEG) is currently the technique of choice for enteral nutrition when these patients are affected with dysphagia (2,3). It is controversial if dysphagia extends survival in ALS. Many times, we have to discuss with patients affected by severe respiratory insufficiency the pro and cons of PEG. But it's unknown if PEG prolongs survival in patients with dysphagia and severe respiratory impairment.
Objectives: To investigate if mild or moderate dysphagia is a predictive factor for survival in ALS patients on continuous non-invasive ventilation (NIV)
Methods: We analyzed retrospectively our database of ALS patients followed from January 1995 to May 2021. Seventy-four ALS patients using non-invasive ventilation (NIV) > 22 h/day were selected, with mild to moderate dysphagia. Patients were grouped according to question 3 of ALSFRS-R (swallowing): group 1, score =4; group 2 (score 2 or 3). Cox proportional hazards regression model with a backward stepwise method was used to identify factors affecting survival. A *p*-value of <0.05 was considered as statistically significant.
Results: We excluded 3 patients who decided to undergo PEG in spite of mild to moderate dysphagia. From 71 ALS patients, 55 (77.5%) were male, median onset age was 61 (1st-3rd IQR, 54–69) and the median disease duration was 12 months (1st-3rd IQR, 6.7–17.2). Spinal-onset was observed in 51 (71.8%), respiratory in 13, (18.3%). Axial onset (7, 9.9%) was less common. The median ALSFRS-R score was 39 (1st-3rd IQR, 34.5–44) at diagnosis. Median total survival was 38.5 months (1st-3rd IQR, 20.2–61.9). Twenty-nine patients (40.8%) had some degree of dysphagia at the time of NIV >22h/day. The hazard of death in these patients was significantly influenced by two predictors, disease duration ($p < 0.0001$) and ALSFRS-R progression rate ($p = 0.0008$) at the time of NIV >22h/day. No other significant predictive factor for survival was found ($p > 0.05$), including dysphagia, gender, age, and onset-form.
Discussion: Dysphagia alone was not a predictor for survival in patients dependent on NIV. Our findings are relevant for giving well-supported information to patients and caregivers when discussing PEG for alleviating fatigue in this group of patients.

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References

1. van Es MA, Hardiman O, Chio A, et al. Amyotrophic lateral sclerosis. Lancet. 2017;390(10107):2084–98.
2. Carbo Perseguer J, Madejon Seiz A, Romero Portales M, et al. Percutaneous endoscopic gastrostomy in patients with amyotrophic lateral sclerosis: mortality and complications. Neurologia (Engl Ed). 2019;34(9):582–8.
3. Castanheira A, Swash M, De Carvalho M. Percutaneous gastrostomy in amyotrophic lateral sclerosis: a review. Amyotroph Lateral Scler Frontotemporal Degener. 2021:1–14.

CMS-47 Palliative care for ALS/MND patients: the collaboration with neurology across Europe and the UK

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Background: Palliative care services have been involved in the care of people with ALS/MND from the 1960s and a survey in the UK in 2000 showed that over 75% of specialist palliative care services were involved in ALS/MND care, but often only in the later stages (1). A survey in 2019 across Europe showed that the collaboration between palliative care and neurology was greatest for ALS/MND and cerebral tumour (2).
Methods: Two online surveys were undertaken of palliative care involvement and experiences of collaboration with neurology across Europe and in the UK in 2020. Palliative care participants were recruited through palliative care organisations.
Results: 126 people, from 11 European countries, primarily from Italy and Switzerland, completed the survey across Europe and 86 specialists completed the UK survey, representing about 40% of specialist palliative care units in the UK. In Europe multidisciplinary team (MDT) working was reported but occupational therapy (27%), speech and language therapy (22%) and spiritual care (41%) were less represented. In the UK MDTs rarely involved speech and language therapy (20%) or dietitians (32%). In Europe 94% reported seeing people with ALS, but this was often late in the disease progression with 40% only becoming involved when patients were at the end of life or the terminal phase. In the UK 45% of services were seeing patients at or soon after diagnosis, 79% according to specific need and only 4% were only at the end of life. The barriers to collaboration were similar in both areas. In Europe 79% collaborated with neurology but there was reluctance of neurology to refer (42%), financial or resource issues (20%) and patient or family reluctance to see palliative care (17%). In the UK 86% collaborated with neurology services, with 60% being part of the MDT, and reluctance of neurologists to refer was rarer (16%) and family/patients' reluctance was uncommon (13%). 41% were planning further collaboration and 95% felt that collaboration was helpful in-patient care. Collaboration with other specialties was common, in particular respiratory medicine (64%) and MND Association (41%).
Discussion: Palliative care services are involved in the care of people with ALS across Europe, but often only in the later stages of disease progression, despite guidelines recommending earlier involvement. In the UK there would appear to be earlier involvement, although the respondents may represent those most involved in ALS/MND care and may not be representative of the whole of the UK. The involvement of MDTs is common, although access to speech and language therapy and dietary advice, may be unusual, which may disadvantage patients with ALS/ MND

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References

1. Oliver D, Webb S. Palliat Med. 2000;14:47–8.
2. Oliver D, Borasio GD, Veronese S, et al. BMJ Support Palliat Care. 2020. doi 10.1136/bmjspcare-2020-002322

CMS-48 Salivary gland radiotherapy for sialorrhea treatment in amyotrophic lateral sclerosis patients: a real word study in 212 patients

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Introduction: This study was designed to evaluate the efficacy and safety of radiotherapy (RT) of the primary salivary glands in ALS patients with refractory sialorrhea.
Materials and methods: The patients refractory to conventional sialorrhea drug treatments treated in our institution between 2010 and 2020 were retrospectively analyzed. RT was delivered by a conventional LINAC, with two opposite beams including the two submandibular glands and two thirds of the two parotid glands before 2014, and from 2015 two lateral beams including the submaxillary glands, and two opposite oblique beams directed to each parotid gland, with reduced oral cavity irradiation. The total dose delivered was 10 Gy in 2 fractions or 20 Gy in 4 fractions. RT efficacy was assessed using the 9-item Sialorrhea Scoring Scale (SSS).
Results: 212 patients were included, and 254 treatments plans (42 reirradiations). Median age was 68 years (range: 38–95), 118 women (56%) and 94 men (44%). Median interval between the first and second radiotherapy course was 7.5 months (range: 1–37). The median SSS score before radiation therapy was 8 (range: 6–9), and 2 at 1 month post irradiation (range: 1–6). There was no > grade 1 toxicities.
At the end of RT, all but 1 patient had an improvement in SSS score: 246 had a complete response (CR) (96%, SSS 1–3) and 7 had a partial response (PR) (3%, SSS 4–5); one patient had a stable SSS score (<1%). CR 1 month after RT was achieved in 96% of the 212 patients who underwent a primary RT, and 100% of the 42 patients who received re-irradiation. Patients treated with 20 Gy versus 10 Gy during the first RT treatment were more likely to have a CR at 1 month (99% vs. 91%, $p = 0.01$), and had a greater decrease in SSS

score (mean difference −6 vs. −5 points, $p < 0.001$). Also, the 42 patients who received reirradiation were more frequently previously treated with the RT 10 Gy protocol (23/42 patients, 55%) versus 57/172 (33%) in patients who did not require reirradiation ($p < 0.01$). The 3-beam irradiation technique, used from 2014 and beyond, did not significantly improve treatment efficacy (reduction of the SSS score by −5.7 points on average versus -5.6 points previously, $p = 0.5$). Nevertheless, it reduced 1 month toxicity, in particular decreased saliva thickening discomfort ($p < 0.001$).
Conclusion: A 20 Gy irradiation in 4 fractions is an effective treatment for ALS patients with sialorrhea, with minimal toxicity, particularly by separately targeting parotid and submaxillary glands.
A shorter RT (10 Gy in 2 fractions) may be proposed in patients with poor medical condition, subject to its lower efficacy and the greater need for reirradiation.

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CMS-49 Dyspnoea of unusual origin in an ALS patient under riluzole: a case study

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Background: Dyspnoea in ALS is primarily due to respiratory failure, more rarely to pneumonia, or pulmonary embolism. Riluzole, an antiglutamatergic drug for ALS, is usually safe. Adverse effects, most frequenty nausea, asthenia, and elevated liver enzymes, are generally mild and reversible. Some patients experience vomiting, diarrhoea, anorexia, dizziness, or rare complications (1).
Objectives: To report on an unusual cause of dyspnoea in an ALS patient under riluzole
Case report: A 81-year-old woman was diagnosed with sporadic ALS in March 2021, and riluzole (Rilutek®) started. The disease was classified as clinically probable, laboratory supported ALS. Eight months before, the patient firstly noted progressive weakness of her right arm, 3 months later on the left, and accidental weight loss of 5 kg. She had no relevant comorbidity, and been vaccinated for Covid-19. At the first appointment in April, ALSFRS-R was 44/48, and Riluzole 50 mg twice daily well tolerated. Neurological examination revealed severe atrophic paresis in upper extremities, mild spasticity in both legs, no bulbar or respiratory signs. In May, the patient complained of decline of weight, hand force, and mild gait disturbance. Paresis of the right anterior tibial muscle, detoriated arm weakness were detected, no disturbance of sniff, cough, speech, swallowing (ALSFRS-R 44/48). In June, she noticed a moderatly progressive exertion dyspnoea, orthopnoea, and mild speech diffculties, no thoracic pain, coughing, chokeing, symptoms of infection. On July, respiratory insufficiency was suspected in pneumological evaluation. Spiromety was normal, diffusion capacity severly reduced. Arterial blood gas analysis showed mild partial respiratory insufficiency with reduced carbon dioxide. As hypocapnia is unusual in ALS, further tests were performed. A thoracic CT revealed reticular changes of the whole lung, except subpleural regions, no signs of infection, nor embolism. Bronchoscopy was unremarkable. Cytology /BAL showed lymphocytosis (34%), reduced CD4/CD8-Ratio (0.15), and a negative multiplex PCR. Interstitial pneumonia induced by riluzole was diagnosed, the drug discontinued, prednison orally 40 mg daily administered. Two weeks later, marked improvement of dyspnea, vital capacity by 300 ml (89%), and diffusion capacity (from 25% to 47%) was found. Prednison was well tolerated, tapering to 20 mg, and follow up in 3 weeks planned.
Conclusions: Interstitial pneumonia (IP) is a rare side effect of riluzole. Only a few cases have been published with an estimated incidence of 0.1% in Japan, 4.3% in one study (2). In our ALS clinic this is the first case since 2000. The diagnosis might be underestimated. It should be considered especially in patients with hypocapnia under riluzole intake, and further work-up (chemical analyses, chest X-ray/CT, pneumological tests) be performed.

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References

1. Miller RG, Mitchell JD, Moore DH. Cochrane Database Syst Rev. 2012(3).
2. Inoue-Shibui A, Kato M, Suzuki N, et al. BMC Neurol. 2019; 19(1):72.

CMS-50 The end of life: a description of the perimortem period in Swedish ALS patients

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Introduction: The clinical course of ALS is highly variable. However, the disease most often generalizes to widespread motor dysfunction, eventually resulting in feeding difficulties, respiratory failure, and death (1). There is currently no effective treatment and the expected survival from onset is short. For the patient and their families, receiving a diagnosis of ALS is a devastating event that prompts reevaluation of life plans and priorities. For many, given the fatal outcome of the disease, a major preoccupation and cause of anxiety is about end of life (2). There is currently little research describing the perimortem period in detail and such a study can hopefully mitigate the ideas about an agonizing end of life.
Objective: To provide a detailed description of the perimortem period in the Stockholm ALS cohort.
Methods: This retrospective study will recruit an approximate number of 100–200 patients who received an ALS-diagnosis between January 2016 and February 2020, at the Karolinska ALS center in Stockholm. Patients who met the revised El Escorial criteria for definite, probable and probable laboratory-supported ALS will be included. Patients have been followed every 3 months during the entire ALS disease. At the time of death several variables were collected to assess the perimortem period and will be used for this study, supplemented by patient medical record data. The study will be describing a wide range of relevant variables such as place of stay (e.g. institution or own home), the use of medications, cause of death, the presence of gastrostomy and/or invasive ventilation. Moreover, these data will be correlated with our extensive follow-up data set specifying clinical disease progression as well as other parameters.
Results: Results will be analyzed presented at the international symposium on ALS/MND in December 2021.

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References

1. Brown RH, Jr., Al-Chalabi A. Amyotrophic lateral sclerosis. N Engl J Med. 2017;377:1602.
2. Averill AJ, Kasarskis EJ, Segerstrom SC. Psychological health in patients with amyotrophic lateral sclerosis. Amyotroph Lateral Scler. 2007;8:243–54.

CMS-51 The clinical saliva score for MND: a validated tool for monitoring saliva symptoms

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Background: Saliva symptoms are common in people with MND, but at present are poorly managed. To assess the performance of potential therapies in future trials, a validated tool to estimate symptom severity is needed.
Objectives: We sought to further develop and validate the clinical saliva score for MND (CSS-MND) (1) such that it could be used to estimate the severity of saliva symptoms in people with MND. The amended CSS-MND is a ten-item patient reported outcome measure.
Methods: Participants completed the CSS-MND, ALSFRS-R, a global change questionnaire and modified Likert scale at 1–5 approximately 3-monthly visits. Those reporting no change on the global question additionally repeated the questionnaires approximately 1 week after visit 1. The construct validity of the CSS-MND was assessed using known groups validity by comparing the mean scores at visit 1 between those with bulbar and limb onset using a *t*-test. The convergent and discriminant validity of the CSS-MND was examined by the correlation between the Likert scale and the ALSFRS-R. The test-retest reliability of the CSS-MND was assessed by the intraclass correlation coefficient (ICC) for the group who reported no change on the global question.
Results: We recruited 504 participants between February 2018 and September 2020. At visit 1, the mean CSS-MND score for those with bulbar onset was 13.8 ($n = 108$) and for limb onset was 5.4 ($n = 275$) (mean difference 8.5, 95% CI: 7.1, 9.9). In addition, the correlation between ALSFRS-R and CSS-MND was −0.8 and between the Likert scale and CSS-MND was 0.7. These data support the construct validity of the instrument. The test-retest reliability of the CSS-MND was high at 0.89 (95% CI: 0.85, 0.92) supporting the reliability of the instrument. The mean change in score between visits 1 and 2 were: 'worse', −2.3 ($n = 95$, SD 5.7), 'about the same', 0.0 ($n = 185$, SD 3.5) and 'better' , +0.5 ($n = 19$, SD 5.9), with a positive mean change suggesting an improvement in saliva problems.
Discussion: The CSS-MND is a tool developed specifically for use in monitoring saliva symptoms in people with MND. We have validated this patient reported outcome measure and further estimated clinically relevant score changes. These could be included as outcome measures in future interventional trials that are badly needed to address the poor management of saliva problems in this patient group.

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Reference

1. McGeachan A, Hobson E, Shaw P, et al. Amyotroph Lateral Scler Front Degener. 2015;16(1–2):108–13.

CMS-52 Information needs and preferences in ALS patients in the Netherlands

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Background: Providing adequate information to patients with Amyotrophic Lateral Sclerosis (ALS), Progressive Spinal Muscular Atrophy (PSMA) and Primary Lateral Sclerosis (PLS) and their informal caregivers is pivotal for making informed decisions in their treatment (1,2).
Objective: To evaluate the information needs of patients with ALS, PSMA and PLS and their satisfaction with the received information with respect to the content, timing, format of information.
Methods: A digital cross-sectional survey has been sent by e-mail or post to a random sample of patients with ALS, PSMA and PLS registered in the Biobank Neuromuscular Diseases of the UMC Utrecht and the Prospective ALS study Netherlands ($N = 270$). The survey contained questions on content of different websites of e.g. the ALS Centre Netherlands (Dutch expert centre for ALS, PSMA and PLS), patients associations, ALS/MND association (the Netherlands), and other disease-specific sources of information such as flyers and oral information given by health professionals. Furthermore the survey contained questions on, timing and preferred format of information communication (oral, written, online).
Results: In total 120 patients (44%) completed the survey. Two-third of the respondents ($N = 79$, 66%) uses the internet as source of information. Main source used is https://als-centrum.nl, the ALS Centre Netherlands. Respondents were generally satisfied with the different sources. Some respondents however experienced a lack of information on new or alternative treatment options and active clinical trials and studies investigating new medication. Furthermore some respondents indicated that they missed the opportunity to ask follow-up questions about received health information, especially in the beginning of the disease trajectory. Of the respondents, 23% ($N = 28$) lacked detailed information on the roles of the different healthcare professionals within the multidisciplinary teams. Information given right after the diagnosis is experienced as confronting but suitable in time. Preferences for the format of written information differed: 58% ($N = 69$) preferred online information, 28% ($N = 33$) written on paper and 15% ($N = 18$) had no preference.
Discussion: The majority of respondents are generally satisfied with the information provision by the different providers within the ALS care in the Netherlands with respect to content and

timing. Patients need customized and adequately timed health information. Websites are an important source of information, healthcare professionals should allocate resources to provide accurate information that is easy to find.

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References

1. Abdulla S, Vielhaber S, Machts J, et al. Information needs and information-seeking preferences of ALS patients and their carers. Amyotroph Lateral Scler Frontotemporal Degener. 2014;15:505–12.
2. Helleman J, Van Eenennaam R, Kruitwagen ET, et al. Telehealth as part of specialized ALS care: feasibility and user experiences with "ALS home-monitoring and coaching". Amyotroph Lateral Scler Frontotemporal Degener. 2020; 21(3–4):183–92.

CMS-53 Experience and usability of mechanical insufflation exsufflation and breath stacking in amyotrophic lateral sclerosis

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Objectives: The published evidence on the usability and participant self-reported benefits experienced by ALS Mechanical Insufflation Exsufflation (MI-E) and Breath Stacking (BS) users is limited. Self-reported comfort and efficacy and quality of life have been investigated, however, areas such as the usability, level of assistance, effect on speech volume and the self-reported effect of MI-E and BS on cough strength during a chest infection have not been assessed. This study had one aim, to evaluate the experiences of participants who were prescribed MI-E and BS. The objectives were to assess MI-E and BS usability, to measure self-reported adherence, to evaluate the self-reported effects of MI-E and BS on cough strength and speech volume and to evaluate the self-reported effects of MI-E and BS on cough strength during a chest infection.
Methods: This was a cross sectional study which took place concurrently with a prospective observational cohort study investigating the decline of respiratory measures in ALS. This study took place at a tertiary ALS/Motor Neuron Disease (MND) Multidisciplinary Team (MDT) clinic in Beaumont Hospital Dublin. Beaumont Hospital Medical Ethics Committee and RCSI Research Ethics Committee granted ethical approval (15/62, REC 1150). A 13 item self-administered questionnaire was developed to evaluate the experiences of participants who were prescribed MI-E and BS and addressed experience and usability, self-reported adherence, self-reported effects of MI-E and BS on cough strength and speech volume and self-reported effect of MI-E and BS on cough strength during a chest infection.
Results: The completed questionnaires describe the experiences of 27 participants prescribed BS and/or MI-E. The main findings of the study indicate that MI-E and BS were user friendly; there was a high level of adherence (96.3%, $n = 26$) with MI-E and BS use; 63% ($n = 17$) agreed that MI-E and BS were easy to use. Less than half (48.2%, $n = 13$) agreed that MI-E and BS increased self-reported cough strength. There was a low level of agreement that the devices increased speech volume (29.6%, $n = 8$) and the self-reported increase in cough strength during a chest infection is undetermined.
Conclusions: ALS participants had a positive experience with MI-E and BS use and MI-E and BS are user friendly. Participants are mostly adherent with MI-E and BS and MI-E and BS increase self-reported cough strength in some participants. Self-reported increase in cough strength during a chest infection is undetermined because of a low number of chest infections experienced. MI-E and BS may play a role in increasing self-reported speech volume for some ALS participants.

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CMS-54 The composite measure of arterial blood gases and respiratory symptoms is predictive of pulmonary function tests in amyotrophic lateral sclerosis

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Background: Spirometry is commonly used to monitor respiratory function in ALS. However, its use is poorly accurate in patients with bulbar or cognitive impairment. Arterial blood gas analysis (ABG) correlates moderately with forced vital capacity (FVC) (1).
Objectives: We aimed at investigating if the evaluation of the ALSFRS-r respiratory items (10 and 11) could increase the correlation between ABG and FVC in a cohort of ALS patients.
Methods: We selected the first ABG performed before non-invasive ventilation by 488 ALS patients. We studied the best

combination to predict FVC by evaluating the correlations of different combinations of ABG parameters (carbon dioxide, pCO_2; carbonate, HCO_3^-) and respiratory symptoms (dyspnea and orthopnea were present if ALSFRS-r items 10 and 11 were <4, respectively) with FVC. Patients were grouped into 3 groups according to ABG values (group 1: normal ABG; group 2: either pCO_2 or HCO_3^- increased; group 3: both pCO_2 and HCO_3^- increased), to compare clinical features between patients with and without respiratory symptoms. For a proper comparison, general impairment was evaluated by ALSFRS-r score without the respiratory domain (ALSFRS-r36) and thus, disease progression rate as ΔALSFRS36.

Results: The best combination to predict FVC was: pCO_2 + HCO_3^- + ALSFRS-r item 10 ($R = 0.430$, $p < 0.001$). In all groups patients with dyspnea showed a more severe general impairment, a higher disease progression rate and lower FVC values. Patients with normal ABG complaining of dyspnea had a reduced survival in comparison with patients without dyspnea (0.91 years, IQR 0.46–1.91 vs 1.46 years, IQR 0.89–2.29, $p = 0.002$). Cognitive dysfunction did not influence the complaining of dyspnea (OR 1.009, 95% CI 0.837–1.215, $p = 0.927$). Among all groups patients with normal ABG and dyspnea showed the highest progression rate (fast progressors; ΔALSFRS36 = 0.86, IQR 0.44–1.25); on the other side, patients not complaining of dyspnea despite having a respiratory failure at ABG had the lowest progression rate (slow progressors; ΔALSFRS36 = 0.38, IQR 0.26–0.52).

Discussion: The ability of ABG to predict FVC increases by adding the clinical evaluation of dyspnea. At equal ABG values, the complaining of dyspnea is associated with lower FVC values. The presence of dyspnea differs according to disease phenotypes, being more frequently experienced by patients with a worse motor impairment and a faster disease progression. A close respiratory monitoring should be set up for fast progressors complaining of dyspnea to look for an initial diaphragm weakness and for slow progressors even without dyspnea because they could show a respiratory failure at ABG.

Conclusions: Combining ABG with clinical evaluation of dyspnea improves the ability to assess early respiratory dysfunction in ALS, especially in patients with bulbar or cognitive impairment.

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Reference

1. Manera U, Torrieri MC, Moglia C, et al. JNNP. 2020;91(9): 999–1000.

CMS-55 Development of the OptiCALS nutritional support intervention for people with amyotrophic lateral sclerosis

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Background: Weight loss is common in people with Amyotrophic Lateral Sclerosis (pwALS) and is a predictor of poor outcomes (1). There is encouraging evidence that increasing calorie intake may affect disease progression (2). Significant variability has been identified in the provision of nutritional management of pwALS across the UK (3). There is a need for evidence-based nutrition support interventions to improve the outcomes of pwALS.

Objective: To develop a complex nutrition support intervention to support pwALS to increase their calorie intake.

Methods and Results: A Portal Development Group (PDG), including academics, healthcare professionals (HCPs), web-developers and public involvement representation were tasked with developing an online portal to provide a personalised experience to pwALS including presenting individualised feedback on calorie intake and weight, and information on nutrition support strategies.

The online nutritional analysis software, myfood24, was integrated into the portal, allowing an individuals' calorie intake to be presented in real time. Intervention content was based on systematic reviews and interviews with pwALS, carers and HCPs identifying the key enablers and barriers to increasing calorie intake in pwALS, using the COM-B model as an overarching theoretical framework. These were then targeted by specific behaviour change techniques in the intervention. The intervention was developed through a series of 'think aloud' interviews with pwALS, carers and HCPs, across six iterations. The PDG made changes to the intervention between each iteration, in response to the feedback received. The online portal was further developed with three rounds of user testing, involving pwALS and their carers engaging with the portal for one month following being trained by HCPs. All participants and HCPs taking part in the user testing were interviewed, with their feedback being used to further refine the portal and training. The final portal, named OptiCALS, is now being evaluated in a multi-centre randomised controlled trial.

Discussion: The development of an online complex nutritional intervention requires significant time and resources. An iterative process of 'develop-test-listen-refine-repeat' involving effective communication with key stakeholders at all stages, is essential in helping to establish an intervention's acceptability and feasibility.

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References

1. Desport J, Preux P, Truong T, et al. Nutritional status is a prognostic factor for survival in ALS patients. Neurology. 1999;53:1059–63.
2. Dupuis L, Oudart H, René F, et al. Evidence for defective energy homeostasis in amyotrophic lateral sclerosis: benefit of a high-energy diet in a transgenic mouse model. Proc Natl Acad Sci USA. 2004;101(30):11159–64.
3. Halliday V, Zarotti N, Coates E, et al. Delivery of nutritional management services to people with amyotrophic lateral sclerosis (ALS). Amyotroph Lateral Scler Frontotemporal Degener. 2021;1–10.