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Theme 13 Clinical management and support

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Theme 13 Clinical management and support

CMS-01 Genetic testing for familial amyotrophic lateral sclerosis (ALS): insights and challenges

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Keywords: genetic counselling, FALS, genetic testing

Background: Pathogenic variants in ALS genes are known to be present in up to 70% of familial and 10% of apparently sporadic ALS cases, and can be associated with risks for ALS only, or risks for other neurodegenerative diseases (eg. frontotemporal dementia). While there are no changes to medical management for patients confirmed as pathogenic variant carriers, genetic testing may be important for future drug trials. Confirmation of a pathogenic variant also provides relatives with the opportunity to consider predictive and/or reproductive genetic testing. Genetic counselling is an important aspect of testing decision-making as it enables individuals to make informed decisions about genetic testing while minimising adverse psychological, ethical and legal outcomes. Few studies have explored how individuals decide whether to pursue testing, nor the needs and experiences of familial ALS families.

Objective: To identify factors that influence patient and family member decision-making about genetic testing for ALS genes, assess the impact of familial disease on the patient and their family, and identify information and support needs.

Methods: In-depth, semi-structured interviews with individuals from Australian ALS families with known pathogenic gene variants explored experiences of familial ALS, and factors that influenced genetic testing decision-making. Interviews were analysed using an inductive approach.

Results: Thirty-four individuals from 24 families were interviewed and included patients ($n=4$), spouses ($n=4$), and asymptomatic at-risk relatives ($n=26$). Life stage, experience of disease, costs, research opportunities, and attitudes to familial ALS and/or reproductive options influenced decision-making. Some patients and relatives experienced difficulty gaining accurate information from their health professionals about the costs and implications of genetic counselling or testing, resulting in a reluctance to proceed.

Discussion and conclusion: This study provides new insight into the Australian experience of genetic testing and counselling for familial ALS. It highlights the need to work together with other health professionals to ensure the complexities of genetic testing decision-making, and referral pathways are better understood.

Acknowledgments: This study was supported by the 2017 Motor Neurone Disease Research Institute of Australia Graham Lang Memorial grant-in-aid. We would like to thank the participants who took part in this study.

CMS-02 Preventing amyotrophic lateral sclerosis (ALS) through reproductive genetic testing: costs and complexities

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Keywords: genetic counselling, pre-symptomatic, prevention

Background: Reducing incidence of ALS is only currently possible in families with a known pathogenic ALS gene variant, through reproductive options that prevent variants being inherited. Options include pre-implantation genetic diagnosis (PGD, through IVF) and prenatal diagnosis (PND, with

a view to terminate an affected pregnancy). Individuals may also choose not to confirm their carrier status and access PGD or PND by exclusion or non-disclosure. Care of individuals deciding about these options relies on decision-making that aligns with their personal values, through genetic counselling. The goal of genetic counselling is not to reduce inherited disease prevalence but to promote informed choices and increase personal control, whilst minimising psychological distress. Uptake of genetic counselling, predictive and reproductive genetic testing is low amongst familial ALS (FALS) families. In addition, certain tests can be costly to individuals (eg. PGD).

Objective: 1. Explore experiences and decision-making about reproductive options. 2. Compare the costs and outcomes of providing funding for ALS families to access reproductive genetic testing options that may reduce incidence of ALS in future generations.

Methods: As part of a wider Australian study exploring individuals from ALS families' choices about genetic testing, qualitative in-depth semi-structured interviews explored experiences of reproductive genetic testing and attitudes and perceived barriers to each option. Interviews were analysed using an inductive approach. The costs of reproductive choices, genetic counselling and testing were analysed with a range of outcomes modelled and compared to reported costs of an ALS diagnosis.

Results: Thirty-four individuals from 24 families were interviewed and included patients ($n=4$), spouses ($n=4$), and asymptomatic at-risk relatives ($n=26$). Unique factors influenced decision-making about reproductive testing options, including experience of disease, religious or moral beliefs, costs, and psychological impacts. We modelled choices and costs relating to 1) predictive testing and genetic counselling 2) reproductive options (PGD, PND, no children, and natural conception) and 3) the probability that offspring of patients with familial ALS inherit pathogenic variants. Modelling health care costs is complicated by the wide range of ages at diagnosis in ALS families and variable penetrance, and is therefore very sensitive to the discount rate used.

Discussion and conclusion: In the absence of a curative treatment for ALS, reproductive genetic testing can provide a cost-effective way to reduce the incidence of FALS. While individuals' choices for testing are not made on costs alone, having information about the costs and barriers to testing informs policy funding decisions and empowers people from MND families to make informed choices.

Acknowledgments: The Motor Neurone Disease Research Institute of Australia 2017 Graham Lang Memorial grant-in-aid supported this study. We would like to thank the participants who took part in the interviews.

CMS-03 The importance of the reception performed by ABrELA's social service to ELA patients and their families

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Keywords: management

Background: ALS is a disease that has not yet been cured, but much can be done to improve the quality of life of the person with ALS and his or her family. Upon receiving the diagnosis, patients and relatives are, for the most part, anxious and distressed by the total ignorance of the disease and the amount of information they need to deal with from that moment on. Reception is essential to strengthen the relationship between the social work professional and all those involved with ALS (patients, relatives, caregivers and health professionals). It is from there that we will be able to understand how the patient and the family are organizing and also to give information and guidance on how to deal with the disease and carry out the appropriate treatment.

Objective: To offer tools that seek to strengthen, support, relieve, motivate and empower patients and families during the course of the disease, so that they have a better quality of life.

Methods: ALS patients and their relatives attended by the ABrELA social service in person, by telephone or e-mail. The data are recorded in the field journal and, at the end of each month/year, a statistic is generated.

Results: The statistics show that, for each first-time host, there are countless subsequent returns, which shows that the bond created by the host generates trust and security for new calls. The reception of the ABrELA social service, having as basic principle the integral care to the patient and family, taking care of people in all their dimensions (physical, mental, spiritual and social) through personal contact, telephone, and also in our meetings that happen every three (3) months, allows the exchange of experiences and experiences that greatly enrich to all who participate in it.

Discussion and conclusion: Valuing life at every moment, learning to live with new limits and possibilities, knowing their rights, giving new meaning to their existence and, above all, knowing that you can count on the support of ABrELA.

CMS-04 The diagnostic experience in MND; a UK survey of the perspectives of people living with MND

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Keywords: diagnosis, clinician-patient communication

Background: Satisfaction with delivery of the diagnosis is dependent on setting, time spent with the patient, physician's knowledge levels and empathy (1). International guidelines exist to aid delivery of the diagnosis; the NICE MND guideline includes recommendations for the time of the diagnosis (2). An Australian survey explored diagnostic experiences from multiple perspectives and confirmed earlier findings (3). Following publication of 2016 NICE MND guideline (2) it was appropriate to evaluate practice.

Objective: To explore the delivery of a diagnosis of MND from the perspectives of plwMND

Methods: An anonymous national survey of plwMND based on the Australian study (3) hosted on Survey Monkey®. A combination of closed and open questions utilising the SPIKES protocol for communicating bad news (4) elicited participants' experiences. Paper copies were also available.

Results: 94 plwMND accessed the survey, but not all responded to every question. Of respondents, 60% ($n=41$) were Male;73% ($n=51$) were married; 57% ($n=40$) were retired . 98% ($n=62$) received their diagnosis from a neurologist during consultations lasting on average 33 minutes (range 5–70 mins); 67% ($n=41$) regarded this as 'just enough' time. Most respondents (56%; $n=37$) knew little about MND before diagnosis; 59% ($n=36$) received 'just enough' information, but 34% ($n=21$) received too little and 28% ($n=17$) were dissatisfied with it. 94% ($n=58$) received their diagnosis in private, from a knowledgeable consultant, but 26% ($n=16$) were unaccompanied. 63% ($n=37$) rated the neurologist's skills/ability concerning the diagnosis as high, while 37% ($n=22$) rated it low. 62% ($n=38$) agreed the neurologist acted with warmth, care and empathy. 51% ($n=31$) were satisfied with the delivery of the diagnosis; 35% ($n=20$) were dissatisfied. Those who regarded the neurologists' skills as low were more likely to be dissatisfied with delivery of the diagnosis. Information given varied; a majority were informed how the diagnosis had been reached (72%; $n=42$), the degree of certainty of the diagnosis (72%; $n=42$), and the prognosis (53%; $n=31$). Fewer learnt about treatment options (48%; $n=28$), progression (41%; $n=24$), local services (38%; $n=22$) and support groups (21%; $n=12$). Only 7% ($n=4$) were informed about advance care planning.

Discussion and conclusion: Over a third of respondents were dissatisfied with how their diagnosis was given, which is comparable with other studies. A high proportion were given their diagnosis by a neurologist, uninterrupted and in private, though over a quarter were unaccompanied at the time. In terms of adhering to guidelines, there is still work to do to improve the delivery of the diagnosis, including the type and amount of information and support given to plwMND.

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CMS-05 Changes in diurnal and nocturnal activity occurs in ALS/MND patients

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Keywords: activity, circadian rhythm

Background: MND is typically characterised by progressive motor deficits, however it is now accepted that for many patients, this is a disease of the wider brain. Non-motor symptoms are prominent & contribute to disability and the burden of disease (1). Studies show a greater degree of fatigue, excessive day time sleepiness, & significant sleep abnormalities early in the disease, despite intact respiratory function & diaphragmatic control. This suggests that abnormal patterning of activity during the day & night may not occur solely as a consequence of respiratory dysfunction, but may be due to dysfunction of mechanisms of circadian patterning. While circadian rhythm dysfunction is well documented in other neurodegenerative diseases (2), there is less known about circadian rhythm dysfunction in MND.

Objective: As a first step towards understanding the possible role of circadian rhythm dysfunction in MND, our objective was to investigate diurnal & nocturnal activity in MND patients.

Methods: Fifty-four patients with probable or definite MND (Female $n=18$; Male $n=36$) with preserved diaphragmatic & respiratory function & thirty non-neurodegenerative disease healthy control participants were recruited as part of an ongoing research project assessing the impact of altered energy expenditure in MND. The average age of the control & MND group was 53.2 (± 13.09 SD) and 58.98 (± 8.86 SD) respectively. For MND patients, the average total ALSFRS-R score was 38.07 (± 5.17 SD), & average ALSFRS-R respiratory sub-score was 11.39 (± 1.09 SD). Average forced vital capacity (% predicted) was 91.51% (± 14.75 SD). Diurnal & nocturnal activity was determined using a wrist worn ActiGraph Link activity monitor. Daily diurnal & nocturnal activity was averaged across 8 consecutive days. Computerised scoring algorithms were used to estimate information about diurnal & nocturnal behaviour from the activity data.

Results: When compared to a matched control population, MND patients spent a shorter amount of time in an active bout ($p<0.01$ CI: 48.31–58.09), had a decreased number of step counts ($p<0.01$ CI: 58586 -72975), higher percentage of sedentary activity ($p<0.01$ CI: 54.56–60.21), & woke up less often ($p<0.01$ CI: 13.67 – 16.2) over the 8-day period. Time in an active bout, number of step counts & the percentage of sedentary activity correlated with the severity of disease, as indicated by total ALSFRS-R scores ($r=0.42$ $p=0.001$; $r=0.66$ $p=<0.01$; $r=-0.55$ $p=<0.01$; respectively), but not the severity of respiratory dysfunction, as indicated by ALSFRS-R respiratory sub-scores ($r=-0.01$ $p=0.92$; $r=0.02$ $p=0.85$; $r=0.04$ $p=0.74$; respectively).

Discussion and conclusion: Changes in diurnal activity & disruption in some aspects of nocturnal activity occur in MND, as indicated by changes in daily activity & the number of awakenings at night. Future studies will determine whether these results correlate with changes in circadian rhythm.

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CMS-06 A survey of healthcare professionals on the measurement of physical functioning in amyotrophic lateral sclerosis (ALS)/motor neurone disease (MND) and attitudes to development of technology based measurement and monitoring solutions

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Keywords: telemedicine, multidisciplinary care, outcome measurement

Background: Specialised multidisciplinary clinics are recommended for people with ALS/MND, however, disadvantages including burden of travel have been identified. Innovative service models utilising telemedicine have been reported (1), but with limitations in patient assessment. Novel assessment methods are required to maximize telemedicine service delivery and priorities for clinically meaningful assessment need identification. Furthermore, the attitudes of healthcare professionals to developments in telemedicine for ALS/MND care requires investigation.

Objective: The aim of the project was to survey healthcare professionals regarding; 1) the use of and attitudes towards current outcome measurement tools 2) their attitudes towards technology based measurement tools, remote monitoring and telemedicine.

Methods: An online survey for healthcare professionals was designed. Demographics and service descriptors were sought. Information was collected regarding physical functioning assessment methods used and the attitudes of the respondents to these tools. The perspectives of respondents regarding the potential for development of technology based assessment methods was sought. Finally, a questionnaire designed to evaluate the acceptance of healthcare professionals to incorporation of telemedicine was completed (2). The survey was disseminated via email invitation, Twitter and Facebook between May and August 2019.

Results: Forty-five respondents from six countries included neurologists, nurses and therapists working in hospital and community based settings, 50% of whom described themselves as expert clinicians. Only 9.7% reported that ALS/MND specific telemedicine infrastructure was used, while 51.6% indicated that this was not available but they would like to use it. Video conferencing was used by 45%, 16% reported using remote monitoring while 38.7% indicated that they would like it use it if available. The most commonly used assessment method were subjective symptoms (89.7%), weight (79.3%) and the ALSFRS-R (67.9%). 65.5% agreed that current measurement tools are clinically meaningful, while only 38% agreed that they are used consistently across sites. Health professionals identified a wide variety of constructs as the most important variables to measure in their patients.

Discussion and conclusion: A minority of clinicians report using telemedicine in ALS/MND clinical care, while a significant percentage indicate that they would like to use it if available. There is concern about a lack of consistency in outcome measure use across sites. New measurement tools providing consistent and valid data regarding physical status and progression are required in ALS/MND.

Acknowledgments: Funded by the HRB Ireland and RMN under the MRCG-HRB scheme 2018.

Thanks to the respondents

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CMS-07 Motor symptoms and physical activity assessment in amyotrophic lateral sclerosis: A systematic review

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Keywords: activity, muscle

Background: Current studies (1–4) on ALS shows the needing for specific evaluation and intervention approaches, with sensitive and validated instruments for better monitoring, screening and assessment of those patients; considering its heterogeneity as a constant barrier to be overcome in advances of treatment research. There is no consensus about the best strategy to evaluate, follow and investigate the progression of this disease regarding those motor and physical presentations (2). Literature shows different ways of motor symptoms (MS) and physical activity (PA) assessment, but there is no evidence of supremacy, sensitivity or application of those.

Objective: To conduct a literature review on MS and PA in ALS and critically analyse of current methods and instruments for muscular strength, gait and postural balance investigation.

Methods: Amyotrophic Lateral Sclerosis, gait, postural control and muscular strength were employed for disabled descriptors of the Medical Subject Headings (MeSH). Articles from 2009 to January 2019, fully available in the Virtual Health Library, PubMed, and Physiotherapy Evidence Database (PEDro) were used, with no restrictions in terms of number of participants, gender, publication status or language. Duplicate articles, those that doesnt show the specific method of motor evaluation and review studies were excluded.

Results: Twenty-six articles met the criteria out of 119 encountered articles. Specifically, those articles were published from 2016 to 2019 (57%). The studies covered measures of muscular strength (23%), evaluation of other muscular characteristics (4%), gait or postural control assessment (15%); association of muscular strength measurements with other muscular characteristics (35%) and association of muscle strength measurements with gait and postural control evaluation (23%). The muscular strength measurements found were Medical Research Council measures (58%), hand held dynamometer (19%), accurate test of limb isometric strength and Tufts quantitative neuromuscular exam (19%), grip strength (4%). The main muscular characteristics measurements were MUNIX and MUNE protocols (46%), electromyography (18%), others like sonoelastography and ultrasound (36%). For gait and postural control assessment, there were cited clinical scales as Berg Balance Scale (BBS), dynamic gait index (55%); the timed up and go test (18%), gait velocity measures (18%), force platform posturography (9%).

Discussion and conclusion: Assessment of MS and PA in ALS has been based on muscle strength and activation. Few studies detailed sensitivity and applicability of the tests, which can make harder standardized indication and use.

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CMS-08 Are we under dosing our patients with mid-stage disease amyotrophic lateral sclerosis? When "usual" activities becomes rowing across the Atlantic

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Keywords: exercise, mobility

Background: Exercise prescription for individuals with Amyotrophic Lateral Sclerosis (ALS) is difficult because of insufficient evidence for specific guidelines across the continuum of disease. However, there is moderate-level evidence

of moderate-intensity strengthening and cardiovascular exercise in early stage ALS does not adversely affect disease progression (1). It is suggested that exercise during later stages of disease or in individuals with poor functional capacity should be cautious and focus on not taxing the neuromusculoskeletal system (1). So, when a patient in mid-stage ALS decides to row across the Atlantic Ocean in a race, are they taxing their neuromusculoskeletal system?

Objective: We describe a patient with ALS during and after a row event across the Atlantic.

Methods: AA is a 57 year old male (175.3 cm, 75 kilograms), with upper motor neuron predominant ALS (El Escorial), with a disease duration of 18 years. His ALS Functional Rating Scale-Revised (ALSFRS-R) of 36 has been stable for 6 years. He uses a power wheelchair in the community and a single point cane at home for energy conservation.

Training program for event. Rowing machine 4 days per week for 45 minutes and 1 day per week for 90 minutes. Additionally, he performed weight-training activities 3 days/week. He focused on stretching and flexibility daily. He increased his caloric intact to 5000 calories with increased fat and protein. He had an intentional weight increase to 89 kg before the event. He reported a marked increase of clonus during the training.

Event. AA as part of a team of five rowed across the Atlantic Ocean, 3548 miles in 51 days, 11 hours, 57 minutes. He performed reduced shifts of rowing for 1.5 hours and longer rests to accommodate cognitive and physical fatigue during the event.

Results: Post- event. AA was successful in completing the row across the Atlantic. He reports he is overall slower, has more fatigue, and worsening dysarthria after the event, described as worse than pre-training.

Discussion and conclusion: Historically high levels of activity may increase the risk of muscle degeneration and/or motor neuron degeneration (2). However, for some individuals' high level of activity may improve neuronal plasticity, improve cardiovascular fitness similar to the general benefit of exercise. High intensity exercise may benefit the healthy motor neurons. Objectives of exercise should be to facilitate healthy innervation, slow degeneration, maintain cardiovascular health, strength, endurance, but not over-stressing the neuromusculoskeletal system.

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CMS-09 Robot-assisted training using hybrid assistive limb for ALS patients

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Keywords: therapy, exercise

Background: Hybrid assistive limb (HAL, CYBERDYNE Inc.) is the cyborg-type robot, by which a wearer's bodily functions can be improved, supported and enhanced. HAL for medical use (lower limb type) received approval to be manufactured and sold as a medical device by the Ministry of Health, Labour and Welfare, and covered costs by public medical insurance in Japan. However, little is known about the effect of HAL for ALS patients.

Objective: To assess the effect of HAL training on the walking ability in ALS patients.

Methods: Nine patients underwent HAL training (mean age 62.6 ± 12.5) at least 9 days during 4–5 weeks (at least twice in a week). Walking ability (2 min walk and 10 m walk), muscle strength and ALS functional Rating Scale-Revised (ALSFRS-R), forced vital capacity (%FVC) were major outcome. Walking ability were conducted by physical therapists trained to perform standardized assessment procedures. The outcome measures were evaluated before and after HAL training.

Results: Two of 9 ALS patients were evaluated under non-invasive ventilation. The mean 2 min walk before training was 89.0 ± 26.0 m, and was significantly improved up to 102.7 ± 32.2 m ($p = 0.02$). However, 10 m walk (gait speed), ALSFRS-R was 37 ± 4.5, and %FVC 87.1 ± 27.7 had not changed.

Discussion and conclusion: Although this study is a single-arm trial, our findings indicate that HAL training can maintain the walking ability and may lead to slower disease progression in patients with ALS.

CMS-10 The ALS steering wheel: a multidisciplinary approach to evaluating driving in ALS

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Keywords: multidisciplinary care, support, driving

Background: A retrospective review on evaluating driving in patients with ALS at the Montreal Neurological Institute & Hospital (MNI/H) ALS Clinic revealed that patients were inconsistently evaluated and vehicle adaptations were underused.

Objective: The ALS Clinic required a systematic approach to all patients followed. This approach needed to be multidisciplinary, quick and easy to administer, kept within federal and provincial guidelines and/or regulations, and used to establish internal guidelines on when to refer patients for on-road evaluations and/or vehicle adaptations.

Methods: Funding was obtained to evaluate the process and success of establishing a systematic approach to the evaluation of driving in ALS, through a prospective quality improvement (QI) plan. Cutoff points for both prevention and intervention were chosen for each individual assessment part of the evaluation. Evaluation frequency has been established. Each evaluation results in a multidisciplinary team recommendation. This systematic evaluation is called the ALS Steering Wheel.

Results: The QI plan was implemented in September 2017, and driving has been addressed with 77 patients holding an active license. Some patients self-identified a need to cease driving, and voluntarily gave up their license. 3 patients had their license revoked by a health care professional from outside the ALS Clinic. 64 patients have been evaluated using the ALS Steering Wheel. 27 patients have been referred for an on-road evaluation with an occupational therapist, subsidized by the funding. The most common flags for prevention are head/neck rotation and arm reach. The most common flags for intervention are the rapid pace walk and grip strength. Of these patients, 22 passed: 16 had conditions applied (ie. timeframe for re-evaluation, use of automatic vehicle, etc.), and 5 required vehicle adaptations. 5 patients failed the on-road evaluation.

Discussion and conclusion: Since implementation, 100% of patients with an active license are now declaring their diagnosis to the provincial driving authority, and communicating information on changes to driving ability during ALS Clinic appointments. The major topic of discussion raised to date is the wait time for on-road evaluations through the public system. The wait time for a private on-road is two weeks, whereas the referrals made to the public system have a 3–9 month wait period. A strong social-spiritual engagement with driving has also emerged. All patients have equated the ability to drive with autonomy and hope.

In sum, this is an OT-driven initiative with the ultimate goal of promoting and prolonging safe driving in patients living with ALS. The ALS Steering Wheel is simple and easy to administer, with no burden on a single health care professional. Adjustments to the tool (removal of those assessments that have not resulted in clinically meaningful results, and addition of a fatigue/exertion scale) have been identified.

Acknowledgments: ALS Canada provided funding for this project.

CMS-11 Cessation of driving in individuals with amyotrophic lateral sclerosis

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Keywords: QoL, ALSFRS-R, driving

Background: Driving is an important indicator of social mobility, independence, and freedom, but requires visual, motor, and cognitive control and the progressive loss of motor function in Amyotrophic Lateral Sclerosis (ALS) ultimately leads to cessation of driving (1).

Objective: We sought to examine the association between ALS variables related to driving capacity and following the individuals until cessation of driving.

Methods: We assessed 28 individuals with ALS who were still driving. ALS variables included age, gender, ALS Functional Rating Scale-Revised (ALSFRS-R), and cognition via Montreal Cognitive Assessment (MoCA). Driving capacity was assessed using a driving simulation with the Lane Change Task program (LCT) assessing baseline score, and during motor, cognitive, and visual distractions. We tracked the individuals after the LCT assessment until cessation of driving. Using a Cox proportional hazard model after adjusting for age and gender we related driving capacity to the event of cessation of driving. Scores for the LCT were converted to Z scores.

Results: Participants were assessed on driving capacity at 11.8 months (SD 13.2) post-diagnosis, had a mean age of 64.3 (SD 8.6) years, 22 were males, had moderate disease with ALSFRS-R scores of 36.2 (SD 5.8; range 36–48), and had mild cognitive difficulty, with MoCA scores of 24.4 (SD 3.8). Eighteen individuals ceased driving (2 deceased) at 5 months (SD 3.7) after driving assessment with ALSFRS-R scores of 30.2 (SD 8.0; range 17–41). The 10 individuals that continued driving had ALSFRS-R scores of 36.4 (SD 5.1; range 31–47). The LCT task with cognitive distraction was related to cessation, hazard ratio 16.85 (95% CI 1.36 – 208.63; $p = 0.03$). The ALSFRS-R scale was also associated ($p = 0.01$).

Discussion and conclusion: When ALSFRS-R scores indicate moderate motor severity, approximately 18 months from diagnosis patients may be ceasing driving. Cognitive distraction during a driving simulation and the ALSFRS-R score were related to driving cessation. Understanding the respective role of motor and cognitive decline in driving cessation is warranted.

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CMS-12 An investigation into whether board certified neurologists are conscious of supporting continuing employment in their patients

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Keywords: QoL, questionnaire/survey, continuing employment

Background: For patients who have progressive neurological diseases such as ALS, continuing to work is important for QOL, not only financially but also in providing satisfaction from living. However, patients often do not know how to continue working with a disability. Whether their neurologist has an awareness or knowledge about continuing employment has a significant impact on patients.

Objective: To clarify how neurologists, who often become the primary physician for these patients, can become conscious of the need to support continuing employment in their patients.

Methods: We conducted anonymous questionnaire survey of 4753 board certified neurologists. The questionnaire investigated respondents' degree of understanding of patient's working situations, awareness of working support, how they are themselves are involved, the types of jobs which may require their involvement, difficulties with support, and the attributes of respondents.

Results: We obtained 1218 responses (response rate 25.6%). When asked about the 17 different potential sources of advice about patient employment, respondents knew about their local hospital advice department in 76.3%, the intractable disease consultation support center in 49.3%, and the head of the public intractable disease job placement office in 27.6%, but knew of other sources in less than 10%. Neurologists who always asked about the job situation at the first visit comprised 57.5%, but this decreased when patients' symptoms worsened (22.5%), and during routine clinic appointments (5.5). The question of whether to consider employment when determining a treatment plan was always considered by 44.2%, sometimes by 52.4%. During ongoing care, supporting ongoing employment and the approach to work was always considered by 24.3%, sometimes considered by 68.3%. Almost all respondents agreed that the role of the attending physician is important, but in actually implementing the necessary efforts, physicians accounted for only 65.4% versus 97.2% done by social workers.

Many respondents felt they did not have enough time to discuss employment concerns during their busy clinics and that they were unsure about giving advice regarding continued employment without fully understanding the patient's working environment and content.

Discussion and conclusion: Neurologists are aware that they play an important role in their patients' ongoing employment and they are (or are trying) to cope, but they have limited knowledge of employment support, and feel unsure about advising patients in light of this. We confirmed that many neurologists expect social workers to take the lead in these matters. However, it is often the case that physicians must first connect patients with social workers to start the process of providing advice and support. At the time of this survey, we provided a brochure to help furnish respondents with the knowledge necessary for supporting their patients' employment, an approach which may be necessary to continue in future.

Acknowledgments: This study was supported by Grant from the Ministry of Health, Labour and Welfare of Japan

CMS-13 An example of possibility for family members continue working while providing care for ALS/MND patients

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Keywords: management, QoL, working

Background: In Japan's social-welfare system it's not expected that ALS patients who need medical care and their families will engage in economic activities while receiving and providing in-home care. However, if one member of a

family becomes an ALS patient and another member of the family provides care for them, the household risks losing the income of both patient and carer.

Objective: To explore the potential economic activities of ALS patients who need 24-hour medical care and their families.

Methods: An additional interview survey was conducted following the survey Ishijima, Adachi, Kawaguchi and Nakajima (1) reported in Boston. Six subjects were surveyed, five of whom had been in their current home care situation for less than five years, so it was possible to observe changes in their circumstances before and after the establishment of their home-care systems but difficult to investigate the longer-term continuation of these systems. Since many of the subjects of the survey asked what factors are needed to maintain their systems, we conducted additional interviews with ALS patients and their families who worked/have been working for more than 5 years.

Results: One way to maintain their home-care system is for patients and their families to run their own care businesses. Regarding the management of such businesses, the process leading up to their establishment was found to be dependent on "peer-support" information such as that provided by patients' associations and pioneering cases. Subjects reported it was difficult to obtain these information from hospitals or local governments. The following conditions for subjects to successfully maintain their own care businesses were identified:

1. Patients and their families having good business sense; 2. Having altruistic thinking that supports other patients with the same disease; 3. Being able to flexibly switch between thinking of a patient and being a manager; 4. Patients being relatively young

Since subjects also mentioned understanding of the structure of the social-welfare system is indispensable as the first step in establishing a home-care system, we made 2000 copies of a manual with photos and easily understood language that can be used by ALS patients and their families.

Discussion and conclusion: This survey shows that at least in some cases it is possible for ALS patients and their family caregivers to work even if 24-hour care is required, a fact that can contribute to the discussion on how to improve the social welfare system. The manuals we created on social media and received about 1,500 requests from inside Japan and overseas.

Acknowledgments: We would like to thank the research participates in our study.

Funding: This research was supported in part by a grant from the Yuumi Memorial Foundation for Home Health Care

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CMS-14 Information needs and resource preferences in Korean family caregivers of patients with amyotrophic lateral sclerosis

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Keywords: caregiver, questionnaire/survey

Background: ALS patients gradually lose the ability to perform daily life activities and ultimately come to completely depend on others for physical and emotional support. Family caregivers are an essential pillar for ensuring and maintaining the home care of the patient with ALS (1). They are responsible for various tasks such as providing support for daily-life activities and participating in decision-making regarding treatment or care (2). It is important to identify their information needs and source preferences.

Objective: The aim of this study was to explore the information needs and sources preferences of Korean family caregivers of patients with ALS.

Methods: This is a cross-sectional descriptive study. The survey was conducted in a total of 108 family caregivers of ALS patients from an ALS/MND clinic in South Korea. The questionnaire included socio-demographic information, clinical characteristics, information needs, and preferred resources of information.

Results: The mean age of participants was 47.32 ± 11.31 years and 69.4% were female. Regarding the relationship with the patient with ALS, 52.8% were a spouse and 38.9% were adult-child. The majority of caregivers focused on information relevant to the 'symptom management and self-care (3.47 out of 4)', followed by 'diagnosis and treatment (3.25)' and 'social support resources (3.19)' categories. With regard to the 'symptom management and self-care' category, 'emergency situation management skills (3.63)' was the most prominent subcategory. 'Diagnosis and treatment' category was 2nd frequently mentioned, with the most reported subcategory being 'clinical trials (3.63). The majority of participants reported using 'internet searching (89.8%)' when searching for information for ALS patients. Also, participants showed an equally high preference of 'health care professionals' (85.2%). For the usefulness of information source, participants ranked the information from the 'health care professionals' is the most useful (3.94 out of 5), with 'online self-support network (3.59)' ranked second.

Discussion and conclusion: Family caregivers of ALS patients participating in this study demonstrated high levels of information needs, especially in the category of 'symptom management and self-care.' Internet searching is by far the most frequently used information seeking behavior, however, caregivers reported from the information from the health care professional the most useful. Therefore, it is imperative for the health care professional to initiate and support online-based education for caregivers of ALS patients on an ongoing basis. However, the resource of the information from the internet is unclear and requires a higher health literacy level which could have ALS patients and caregivers suffer from inaccurate information. Thus, we suggest building online flat foam for ALS patients and caregivers who can provide reliable and useful information and access to the up-to-date high-quality digital content.

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CMS-15 Aspiring to minimise aspiration: prevalence and predictors of aspiration in progressive neurological disease

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Keywords: dysphagia, video fluoroscopy

Background: The occurrence of aspiration in people diagnosed with Motor Neurone Disease (MND) is high relative to the general population, yet risk factors of aspiration in this population remain understudied.

Objective: The objective of this research project was to identify the prevalence, clinical signs predicting aspiration, and correlation of anatomical features to occurrence of aspiration in patients with a Progressive Neurological Disease (PND) who undergo a Video Fluoroscopic Swallowing Study (VFSS).

Methods: A retrospective cohort study was conducted of patients with a diagnosis of a PND, of which MND was the predominant diagnosis (47%) that undertook a VFSS between 2016 and 2017. Data was extracted from the 175 eligible VFSS recordings. Aspiration was determined by scores of ≥6 across fluid and diet consistencies using the Penetration-Aspiration Scale (PAS) (1). Anatomical domains of the Bethlehem Assessment Scale (BAS) (2) and pharyngeal bolus transit time were examined and correlated with aspiration status. The VFSS were embedded with real time video imaging of patients allowing observation of clinical domains of self-feeding, wet vocal quality, visible distress signs, and uncontrolled movements and their predictive value of aspiration was evaluated using odds ratio.

Results: The overall prevalence of aspiration was 21.7%, with 50% of cases demonstrating silent aspiration. Within the MND population, aspiration prevalence was 28%, compared with 18% aspiration prevalence in other PND diagnoses. Lip and jaw function, pyriform sinus residue, and pharyngeal function correlated consistently with aspiration of regular fluids, mildly thick fluids, and puree, while visible signs of distress significantly predicted aspiration ($p = 0.005$).

Discussion and conclusion: This study provides confirmatory evidence that aspiration prevalence among patients with a PND, and more specifically with MND, is high relative to those individuals with a typical swallowing profile. The number of silent aspiration events highlights the importance of instrumental analysis in assessment of swallowing and prevention of adverse health outcomes such as aspiration pneumonia. This research suggests that aspiration prevalence within the MND population is greater than other PND populations and findings of previous literature. The relationship between anatomical domains and aspiration, in addition to, the significant predictive value of visible distress warrants further investigation into potential clinical bedside predictors of aspiration to identify patients at risk.

Acknowledgments: This research project was a collaboration between Calvary Health Care Bethlehem and the University of Melbourne.

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CMS-16 Providing collars for people with motor neurone disease: trialling not prescribing

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Keywords: assistive technology, physiotherapy

Background: Progression of Motor Neurone Disease (MND) commonly leads to weakness in the neck and trunk muscles. 8.4% of people with MND were found to be using a collar in a recent study (1). Finding acceptable, effective, comfortable, affordable and timely head support can be a challenge. This is especially the case when the person with

MND is still ambulant. Difficulties with swallowing, oral secretions, breathing, and using non-invasive ventilation also complicate this decision. The NICE Guidelines [2016] recommend the provision of equipment and adaptations that meet the person's needs without delay, so that people can participate in activities of daily living and maintain their quality of life as much as possible (2).

Objective: Develop, implement and evaluate a collar-trial service within an MND clinic.

Methods: A range of collars have been procured which can be trialled by people with MND attending the clinic. This includes soft collars, MND/Oxford collar, Headmaster collar, LA wire frame collar, Hensinger collar, Philadelphia collar, Aspen Vista collar, Head Up collar. The collars can also be loaned for short period, to give the person with MND the chance to use the collar at home, while participating in different activities, before deciding if a particular collar meets their needs or not. Data was kept over a six month period of all the people with MND attending SPNDS who trialled collars. Records were kept on which collars they had previously used, if any. Those that were trialled and rejected, and the ones that were deemed to be most suitable for that stage of the disease. Also whether they were still ambulant at the time this decision was made.

Results: The results show that there was a great variety in the collars that individuals choose to purchase, and that needs changed over the period of disease progression.

Discussion and conclusion: The wide variety of collars chosen demonstrates that there is no one 'ideal' collar on the market at present for people with MND. So whenever possible, it is important for people with MND needing a collar for head support should have the opportunity to trial different collar options before deciding on which one will be the most acceptable. The person with MND needs to be an active participant in the decision making process, rather than a therapist prescribing a collar that they think will be suitable or that they are familiar with.

Acknowledgments: The physiotherapy staff at CHCB for collecting this data.

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CMS-17 Clinical implications of upper esophageal sphincter restriction in motor neurone disease

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Keywords: dysphagia, bulbar, High-Resolution Impedance Manometry

Background: Dysphagia in MND is presumed to be due to impaired motor function in the oropharynx. Limited published (1) and historical anecdotal data show temporary improvement in swallow function following botox injection or cricopharyngeal myotomy, related to removing upper esophageal sphincter (UES) restriction.

Objective: We studied swallowing biomechanics using pharyngeal high-resolution impedance manometry in patients with bulbar and pseudobulbar UES impairment to determine biomechanical differences and swallowing outcomes.

Methods: Sixteen MND patients (8M, 70 ± 8yrs) with bulbar involvement were recruited and compared to thirty asymptomatic age-matched controls (C; 14M, 69 ± 11yrs). Patients were classified as bulbar (B) or pseudobulbar (PB) based on neurological examination. All patients and controls underwent high-resolution impedance manometry using 5ml normal saline boluses (MMS Solar, Unisensor catheter with 36 pressure sensors (1cm); 16 impedance segments (2cm)). Data were exported and analysed using Matlab algorithms for interpolated data along the pharynx and at the UES and interpreted as previously described (2). Swallowing function outcomes were clinically and radiologically assessed for aspiration and pharyngeal residue (PAS score (3) and normalized residue ratio index (4)). Comparisons were done with one-way ANOVA with post-hoc Dunn (Bonferroni correction) with p-value <0.05 significant.

Results: Patients with bulbar and pseudobulbar MND had evidence of UES restriction (B 3.7 ± 0.4 mS, PB 4.1 ± 0.3 mS vs C 7 ± 0.5 mS; P <0.001] and pseudobulbar patients also had reduced UES relaxation (UES IRP PB 6.1±2.7mmHg vs. C 0.3±1.1mmHg; P<0.05). UES baseline tone was reduced in both MND groups compared to controls (B 12±4mmHg, PB 35±5mmHg vs. C 55±12mmHg; P<0.001) and was lowest in bulbar, even compared to pseudobulbar (P<0.01). Contractility was similar in all regions above the UES with the exception of tongue-base contractility, which was reduced in pseudobulbar (81±14mmHg vs. 151±17mmHg; P<0.01).

Discussion and conclusion: Overall, MND patients with bulbar impairment (B) had greater UES restriction than controls in the presence of intact tongue base contractility – this combination predisposes them to a greater aspiration risk (than pseudobulbar) due to increased chamber pressures above a more closed sphincter. A degree of intact hypopharyngeal contractility in both B and PB patients implies cricopharyngeus myotomy may be worth considering in both groups for potentially different outcomes (decreasing aspiration risk vs. maintaining oral feeding).

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CMS-18 Interaction between decline of swallowing and cognitive function in motor neurone disease

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Keywords: swallowing, cognitive impairment, dysphagia

Background: Dysphagia (swallowing impairment) occurs in nearly all people with MND as the disease progresses (1). Dysphagia significantly affects patient health through weight loss, dehydration, malnutrition, and respiratory compromise, increasing caregiver burden and reducing quality of life. The MND Australia Research Priorities Survey (2) highlights dysphagia as the 2nd highest research priority for management and care. In addition, nearly half of people with MND experience mild to severe cognitive and/or behavioural changes (3), which are associated with shortened survival, impaired decision making and reduced adherence to treatment strategies. Emerging evidence suggests cognitive changes may impact on a person's ability to recognise and, therefore, manage dysphagia, eg. due to a lack of awareness of swallowing decline or sensorimotor deficits resulting in undetected (silent) aspiration (1,4).

Objective: A scoping review was undertaken to explore and summarise the existing literature using Arskey and O'Malley's (5) methodological framework, and to address the following research question: *How do cognitive and/or behavioural changes impact on a person with MND's ability to recognise, understand and manage dysphagia?*

Methods: Subject headings and relevant keywords were searched across MEDLINE, SCOPUS, CINAHL, PsychINFO, Emcare and Google Scholar databases in May 2019.

Results: Of a total of 805 articles, 16 articles meeting inclusion criteria were included. Full texts were reviewed for current methods of assessing cognitive and/or behavioural change and swallowing function and data grouped by subjective (self-reported)/objective assessments. Key themes exploring the interaction between cognitive and/or behavioural changes, swallowing awareness, and swallowing function over the course of disease progression were extracted.

Discussion and conclusion: Preliminary analyses show that swallowing assessments are conducted frequently as part of routine clinical practice to determine swallowing safety and management strategies. However, cognitive function is not routinely assessed as part of standard speech pathology clinical practice. Thus, the impact of cognitive change on a person's ability to understand, decide on and implement recommendations remains unknown. The outcomes of this review will inform trans-disciplinary care for individuals with dysphagia and contribute to better clinical and quality of life outcomes through holistic, patient-centred care.

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CMS-19 A clinical bulbar assessment scale for ALS (C-BAS)

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Keywords: bulbar, dysphagia, dysphasia

Background: The purpose of the C-BAS is to provide an instrument for comprehensive assessment of bulbar function in patients with amyotrophic lateral sclerosis (ALS), with the broader goal of providing reliable data collection for clinical and research applications. This measure represents a compilation of evidence based clinical components specifically designed to facilitate early recognition and track rate and functional impacts of bulbar motor neuron disease. This instrument differs from commonly used ALS scales by attempting to (a) quantify specific features of speech and swallowing subsystems; (b) maintain sensitivity to

longitudinal change; (c) assess impact of functional impairments on activity and participation. The cumulative data set is projected to accurately identify pre-symptomatic impairments, allowing for timely application of clinical interventions. By expanding the available methods of bulbar assessment through targeted characterization of speech and swallowing, the C-BAS may prove useful for documenting early bulbar impairment and successively monitoring ALS disease progression.

Objective: To develop a valid scale that reliably and accurately identifies symptoms of bulbar dysfunction and tracks the progression of motor neuron impairment over time.

Methods: The C-BAS represents a collection of subjective and objective clinical bulbar measures specifically designed to identify speech and swallowing impairments and track disease progression in ALS. This scale employs quantitative measurement of speech and swallowing, including diadochokinetic rates, maximum phonation time, speaking rate, swallowing rate, sentence intelligibility, the S/Z ratio, reading/pause rate, swallowing tasks and includes patient reported outcome measures. C-BAS provides a profile of UMN and LMN features as well as subjective patient-reported swallowing effort and fatigue, speech effort and fatigue and communication effectiveness ratings.

Results: Cumulative data collection, along with clinical components of the C-BAS will be provided at the time of presentation, which will also include: 1. Content validation data based on a survey of ALS experts reporting on individual C-BAS items; 2. Results from an ALS clinical care C-BAS implementation study ($n = 20+$ individuals); 3. A comparison of C-BAS scores with existing bulbar scales (eg., ALSFRS bulbar subscale, CNS-BFS), in an attempt to achieve validation.

Discussion and conclusion: The C-BAS incorporates a novel means of systematic data collection for assessing early and longitudinal bulbar symptoms. Unlike existing ALS scales, C-BAS is constructed purely for assessment of bulbar dysfunction in a clinical setting. The designed focus is to evaluate impairment of body structures and function (ie., speaking rate, swallow safety, respiration, phonation, affect control), activity limitation (ie., speech intelligibility, swallow efficiency) and participation restrictions (ie., effort, fatigue, eating, interacting, communication effectiveness) not routinely measured through existing ALS instruments. Once validation has been established, this scale should provide a standardized, comprehensive measure targeting bulbar speech and swallowing features in ALS.

CMS-20 Developing a message banking pathway: the Irish experience through multi-agency collaboration

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Keywords: AAC system, speech

Background: Message banking is a Speech and Language therapy (SLT) clinical intervention used to support people with MND (pwMND) to prepare for potential use of an electronic communication aid (ECD) in the future. Message banking involves the pwMND choosing and recording personnally meaningful phrases in their own unique voices, with their own personal delivery and intonation. As defined by Costello (2012) (1), these Recordings 'have an important role both in optimising communication, but also in maintaining a sense of self'. There was previously no standardised Irish message banking pathway. Technical considerations combined with the psychological aspects of planning for the future can cause both the pwMND and the supporting SLT to feel overwhelmed by the process therefore clinical guidelines would help to reduce these daunting elements of this intervention.

Objective: A joint service delivery initiative between SLTs in three Irish agencies was undertaken to create a unified message banking clinical pathway to support both pwMND and their local SLT clinician. The three agencies involved were the neurology SLT service in Beaumont Hospital, Dublin, the national assistive technology service in the Central Remedial Clinic (CRC) and the Clinical Speech and Language Studies department in Trinity College Dublin (TCD).

Methods: Through multiple iterative cycles, an easyread single page pathway document was developed to guide pwMND and SLTs through the key decision making and action points for message banking. Due to an identified clinical need, the related process of voice banking was also included on the pathway. Building on a previous project where focus groups were held with service users regarding their experiences with message banking, the data collected was used to structure the content of handouts. For each decision or action point, a support handout provides detailed information.

Results: A clinical process map was developed to guide decision making and action points, along with resources to support each step at a local level.

Discussion and conclusion: Message banking is a useful clinical tool in supporting pwMND to maintain their identity and social closeness in future ECD use. This collaborative approach facilitated the development of professional guidelines within a national environment of limited resources. By building a team including an acute SLT with extensive clinical experience of working within a

multidiciplinary MND clinic, an SLT working on an assistive technology service that implements voice recordings onto ECDs and an academic SLT with experience in research design and project implementation, we pooled our relative strengths and varied perspectives to build a cohesive professional toolkit. This approach could be applied to other projects and other clinical areas.

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CMS-21 A single adjustment of the scanning-speed of the device affected each participant's feeling and number of letters as an effect of short-term training programs on augmentative and alternative communication to support ALS/MND patients

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Keywords: education, communication, AAC system

Background: Augmentative and alternative communication (AAC) has been used as support for those suffering from neurological diseases, such as ALS/MND patients with communication difficulties. However, patients may not receive enough information and support to adapt to communication aids because medical professionals do not fully understand the necessity on the communication support. AAC has not always been used in appropriate and adequate ways because of insufficient knowledge and skills. We propose the short-term training programs for AAC among undergraduate students in healthcare fields (1). We focused how efficiently a single adjustment of the scanning speed of the electric device affect each participant's feeling and number of letters in the designated time.

Objective: To evaluate each participant's feeling and number of letters in five minutes before and after their adjustments to AAC equipment on the training program for students both within the single session and between the two sessions.

Methods: The training programs, which consisted of lectures and hands-on sessions, were conducted for students at four universities (medical, nursing, rehabilitation or clinical-psychology courses). A total of 18 novice and experienced 23 students participated. In Japanese Scanning Communication Aids (Let's Chat®) (LC), the scanning speeds of the devices were settled in the middle at first (default); then the speed was adjusted as per the preference of each participant (adjustment). It was compared based on the following three points: 1) the number of letters caught in five minutes by LC in default/adjustment, 2) the sense of burden before/default/adjustment by the visual analogue scale, 3) the feelings default/adjustment. The text data were analyzed by a quantitative content analysis. The study was approved by the Ethical Committee of the institutions.

Results: 1) Compared to LC in the default (novices 13.3 letters, experienced 15.1), LC in the adjustment (novices 25.7, experienced 32.4) had a higher number of transmitted characters ($p < 0.01$). 2) The sense of burden, which was low before the experience (novices 15.6, experienced 15.0), increased after the default (novices 39.2, experienced 35.4) and decreased after the adjustment (novices 21.2, experienced 19.0) ($p < 0.01$). 3) The feelings of the participants having excessive expectations from the high technology of LC devices before the experience, showed a sense of difficulty in operation after the experience and the recognition of the importance of adjusting.

Discussion and conclusion: This study reveals two important points. First, the importance of adjusting AAC equipment is confirmed quantitatively. Second, future supporters for ALS/MND patients can recognize the importance of adjustment of AAC equipment based on their own experience and statistical analysis. It is believed that the short-term trainings programs contributed to the learning effect of the participants.

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CMS-22 Gross numbers of letters as the effect of a short-term training program for augmentative and alternative communication on healthcare students after 6 months

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Keywords: education, communication, AAC

Background: Augmentative and alternative communication (AAC) is used to support ALS patients with communication difficulties. However, AAC was not always used in appropriate and adequate ways because of insufficient support (1). Here, we planned a short-term educational training program for increasing AAC skills in students from multiple health-care fields who aim to support patients with ALS (2).

Objective: We aimed to evaluate gross numbers of letters as the effect of the short-term training program on the students' AAC skills after 6 months.

Methods: The training programs were conducted with students from four universities that included at least one medical, nursing, rehabilitation, or clinical psychology course. The training included lectures and hands-on sessions in how to use three types of AAC: ie., a Flick-Type communication board (Flick), Kuchimoji (an oral and eye-blink method without a board), and Japanese scanning communication aids (Let's Chat®: LC). We measured the number of letters obtained in 5 minutes by each AAC and asked about the burden before and after each method using a visual analog scale. We compared the number of letters between the novice group using AAC for the first time and the experienced group who had completed the same training program around 6 months earlier. R (v. 3.5.0) was used for statistical processing. This study was approved by the Research Ethics Committee of the Faculty of Medicine at Mie University (No. 3245, March 2018).

Results: The 18 novice and 23 experienced students participated in the program until June 2019. In the LC session, the devices' scanning speeds were initially set to a middle range and then measured. The speed was then adjusted using each participants' preference and measured again. The novice group's results were: Flick, 30.4 ± 7.2 letters; Kuchimoji, 26.9 ± 9.9 letters; LC, 13.3 ± 2.5 letters; after adjustment, 25.7 ± 4.7 letters. The experienced group's results were: Flick, 35.6 ± 6 letters; Kuchimoji, 33.3 ± 7.4 letters; LC, 15.1 ± 1.7 letters; after adjustment, 32.4 ± 11.4 letters. The number of letters in the experienced group was higher than that in the novice group (Student's *t*-test: Flick, $p = 0.019$; Kuchimoji, $p = 0.027$; LC, $p = 0.01$; after adjustment, $p = 0.045$).

Discussion and conclusion: The results suggested that using AAC made it possible for students to acquire AAC skills and maintain them for 6 months. However, Arcoraci et al. suggested that high-level simulation education could preserve a learning effect for 3 months (3). In this study, the experience of training assumed that the patients contributed to the participants' learning effect.

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CMS-23 Verification of the effect of medical coordinator of intractable disease on the regional medical care network for intractable disease in Japan

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Keywords: management, multidisciplinary care, medical coordinator

Background: In Japan, the medical coordinator (MC) for patients with intractable diseases, such as ALS/MND, plays a role in seamlessly supporting patients from diagnosis to home care. The MC is responsible for maintaining the medical care network for intractable diseases, which includes the medical, health, and welfare staff surrounding patients and their family. In our area, the MC conducts a delivery-type workshop to improve the abilities of health care workers and activate the medical care network in the area. The workshop has been conducted for 10 years since 2008.

Objective: To verify whether the activation of the regional medical care network for intractable disease was achieved after the delivery-type workshop for intractable disease.

Methods: Group interviews were conducted with six people (one director of the regional collaboration office, two medical social workers, one care manager, and two public health nurses) who played central roles in the delivery-type workshop. Six months after the delivery-type workshop for intractable disease, we investigated changes in health care workers and medical care networks for intractable disease in the area. The interview survey was conducted in the consultation room of a hospital for approximately 90 minutes using a semi-structured interview method. The data comprised a verbatim record, extracted related sentences and paragraphs, and code names. To ensure reliability and validity of the analysis results, the results were confirmed by the subject themselves at the code extraction stage. The study was approved by the Ethical Committee of our institution.

Results: Based on the survey results of comparing before and after the delivery-type workshop for intractable disease, significant changes were confirmed in the following three areas. 1) The regional main hospitals changed to conduct meetings for health care workers involved in treating patients with intractable diseases and transmitting information, making a multidisciplinary team, 2) The community health center changed to publicize the health care policy activities for patients with intractable diseases such that the whole community could be visualized, 3) The health care workers treating people with intractable disease in the area began to envision the future of the medical care network.

Discussion and conclusion: This study highlighted two important points. First, with the opportunity for multiple health care workers to meet at workshops and discuss cases, they developed face-to-face relationships. Second, the delivery-type workshop for intractable disease, conducted by the MC and medical experts, activated the medical care network for intractable disease in areas with lower social resources. It is believed that MC activities was useful for the regional medical care network for intractable diseases and improved the quality of support for patients with intractable diseases such as ALS/MND.

CMS-24 Virtual reality in ALS

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Keywords: education, disease burden

Background: Community outreach and engagement are important components in developing increased awareness regarding ALS and motor neuron disease. Patients with ALS (PALS) have demonstrated desire to participate in fostering education regarding ALS for the community and health care professionals. Virtual reality (VR) is a novel means to utilize developing technology to raise public awareness and provide education about motor neuron disease. Virtual reality can create an embodiment of ALS that helps participants develop a deeper understanding of the disease process.

Objective: Develop virtual reality scenarios with current PALS to help educate the community and other health care professionals regarding ALS.

Methods: A partnership was developed with the Virginia Commonwealth University School of Medicine and the School of Arts in creating three VR scenarios involving ALS. Three unique VR scenarios were developed from different PALS with varying societal and educational backgrounds at varying stages of the disease process. The VR embodiment scenarios were developed through two to three one hour interview sessions over a two month period. The total interview time per PALS was approximately five hours.

Results: The creation of a VR scenario in ALS is feasible and PALS found support and positive engagement through the process. Dissemination of the VR scenarios is currently underway within the community. Future research will examine the impact VR may have in the community in addition to improving empathy and humanistic qualities in students, residents, health care professionals, and staff.

Discussion and conclusion: VR is an emerging technological advancement that has the ability to augment other teaching methodologies and has been well received throughout society. Although the utilization of VR in medicine is yet in its' infancy, it is gaining increasing attention as a powerful tool and is being well received by those engaging with it within the field and public. VR provides a new strategy in helping to educate the community about ALS and has the ability to become an inimitable teaching tool throughout the health care community. Further work is ongoing in attempts to quantify the impact on education of health care professionals.

Acknowledgments: We would like to acknowledge the Harper's Hope Fund for ALS at the Medical College of Virginia Foundation which helped to financially support this work. We would like to acknowledge our patient and their families for their commitment to bringing awareness of the disease.

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CMS-25 Improved survival in amyotrophic lateral sclerosis patients following intrathecal administration of autologous bone marrow mononuclear cells (BMMNCs)

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Keywords: survival, stem cells

Background: Despite improved understanding of the disease pathology, current standard treatment is unable to satisfactorily modify the underlying disease process. The unmet medical need and positive pre-clinical and clinical data has highlighted stem cell therapy as a promising avenue in the management of the disease. Our previous study demonstrated beneficial effect of cell therapy on survival duration (pilot-study).

Objective: The aim of our study was to evaluate the effect of combination of intrathecal administration of BMMNCs and short-term lithium along with standard treatment on survival duration in a larger population.

Methods: This controlled study included total 213 patients with probable or definite ALS on Revised El-Escorial criteria. Treatment group ($n=162$) received autologous BMMNCs intrathecally with short term lithium while patients that did not receive cell therapy and/or lithium formed control group ($n=51$). Survival duration from onset of disease was estimated and compared using Kaplan-Meier survival analysis. Also, within the treatment group, the effect of onset type, gender and hormonal status on survival duration was studied.

Results: Mean age of disease onset was 48.41 ± 10.74 (24–76) years in treatment group and 54.17 ± 8.21 (32–77) years in control group. Of 56 females in treatment group 21 were premenopausal and 35 were postmenopausal. Of 106 males, 48 were ≤40 years of age at disease onset and 114 males were >40 years of age at disease onset. No major adverse-events noted. Mean estimated survival duration was 101.5 months in treatment group while in control group it was 47.9 months ($p=0.007$). Survival duration was higher in patients with limb onset (104.5 months) as compared to patients with bulbar onset (63.1 months) ($p=0.059$). Survival duration was significantly higher ($p=0.002$) in premenopausal women (86.7 months) as compared with postmenopausal women (61.2 months); and in males with disease onset at ≤40 years (209.9 months) compared to >40 years of age (57.6 months) ($p=0.000$). Testosterone levels decline after age of 40 years hence it was chosen for studying the effect of hormones on survival duration in males.

Discussion and conclusion: BMMNCs provide immunomodulation thus detoxifying the micro-environment (1). Lithium has shown to demonstrate neuroprotective effects in animal models (2). The results of our study show that autologous BMMNCs are safe and improve survival duration in ALS. Also, younger male patients, limb onset ALS and pre-menopausal females demonstrated better treatment efficacy. The neuroprotective role of sex hormones in ALS should be further explored.

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CMS-26 The effect of Riluzole oral film on swallowing safety in individuals with amyotrophic lateral sclerosis

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Keywords: riluzole, dysphagia, swallowing

Background: Riluzole is a benzothiazole derivative with neuroprotective effects and survival benefit in patients with ALS. Tablet administration of riluzole can be a challenge given that most ALS patients develop dysphagia during disease progression impacting their ability to take medications by mouth. Riluzole oral film (ROF) is an alternate dosage form that adheres to the tongue with the active ingredient in a polymer-based film matrix utilizing Aquestive Therapeutics' PharmFilm® technology. This technology removes the need to transport a pill from the oral cavity to the stomach and to take medication with water, potentially reducing burden on patients with impairments in swallowing efficiency (bolus propulsion) or swallowing safety (aspiration of liquids or pill). Riluzole is known to cause circumoral paresthesia. The effect of riluzole on swallowing safety has never been studied.

Objective: Assess the impact of administration of a single dose of 50mg ROF on swallowing safety in individuals with ALS.

Methods: This study represents a single-center, open label, phase II trial in nine individuals with ALS. Inclusion criteria were: age 18–80, probable or definite ALS (Revised El-Escorial Criteria), an oral diet consistent with a Functional Oral Intake Scale >3, and no known allergy to barium sulfate, riluzole or inactive ingredients in ROF. After consent, participants underwent a standardized Videofluoroscopic Swallowing Study (VFSS) consisting of ten bolus trials of foods and liquids. Subjects were then given a dose of ROF 50mg, on the median sulcus of the tongue. After three minutes, the VFSS protocol was re-administered for comparison of swallowing safety before and after ROF. Swallowing safety was assessed utilizing the validated Penetration Aspiration Scale (PAS) by two independent raters blinded to test condition. PAS scores were presented in multiple formats: individual bolus scores, sum of scores, single worst score, and Steele's 4-level category (based on 8-point original PAS score). Given the small sample size all analysis was descriptive, with comparison within each subject and across subjects.

Results: 55.6% of subjects were able to swallow safely by PAS standards both pre- and post-dose. One patient moved from the "safe" to "penetration" category. No patient had a score indicative of aspiration either pre- or post-dose. There was no evidence of deterioration of swallowing function post-dose, whether analyzing PAS by single worst score, sum of scores, or Steele's 4-level category methods. There were no adverse events reported.

Discussion and conclusion: A single dose of 50 mg ROF had no detrimental effect, was well tolerated, and may

represent an alternative administration route in individuals with dysphagia.

Acknowledgments: We would like to thank patients for taking part in the study. Funding for this study was provided by Aquestive Therapeutics. CB, CJ, and GS are employees of Aquestive. AHH served as a paid consultant to Aquestive.

CMS-27 Nursing driven initiative to increase tolerability and compliance of BHV-0223 Novel Therapy in ALS patients

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Keywords: clinical care

BHV-0223 is a novel, disintegrating tablet formulation of Riluzole designed for sublingual (SL) administration to optimize pre-gastric absorption and bypass first-pass metabolism. Protocol allows treatment with BHV-0223 to eligible adults who have Amyotrophic Lateral Sclerosis prior to it being available on the market.

BHV-0223 40 mg Zydis sublingual formulation is bioequivalent to Rilutek 50 mg tablet swallowed with water under fasting conditions (1).

Eligible patients receive one 40 mg of BHV-0223 (compared to 50 mg of oral Riluzole), administered sublingually, once daily basis for the first seven days of treatment and twice daily basis (approximately every 12 hours) after the first week. As Riluzole tablets are a well-established therapy, standard of care patient education is provided.

Reported incidences of tolerability from patients for adverse event collection purposes revealed several occurrences of poor drug tolerability due to undesirable sensations in the mouth, throat, and neck post sublingual administration of BHV-0223. All patients reported some degree of oral hypoaesthesia and/or numbness after administration of sublingual tablet. Tongue and lips are most typically affected. While the duration of these sensations vary, none last more than 45 minutes in duration, and intensity tends to decrease over time. Tolerability issues related to these findings contributed to discontinuation of drug in 6 of 30 patients, an incidence of approximately 20%.

Given the route and unique findings related to BHV-0223 investigational product, it was clear that additional nursing support was needed to improve the patient experience, and thus compliance, with this drug. Few proven therapeutic agents are available and standard in the treatment of ALS. Therefore, it is crucial that we support our patients as they begin novel therapies in order to maintain compliance. From this experience, a nursing driven education initiative was put into place on the day of initial BHV-0223 dispensation, aimed to better prepare patients for side effects and increase tolerance of this form of Riluzole.

The nursing driven initiative consisted of educational support provided to patient and caregiver at the time of drug dispensation, including one on one counseling with an RN experienced in the administration of BHV-0223. Side effect profiles and incidents of oral hypoastheia are explained to provide anticipatory guidance to aid the patient through any oral numbness or sensation that may be deemed noxious or lead to discontinuation of this form of riluzole. Implementation of the initiative will be assessed to determine if providing anticipatory education on tolerability profile of BHV-0223 will improve drug adherence for this novel therapy.

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CMS-28 Zydis Riluzole 40mg oral disintegrating tablet biohaven expanded access program – single center experience

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Keywords: riluzole, therapy

Background: BHV-0223 is a novel, sublingually administered disintegrating tablet. Formulated to increase pre-gastric absorption and bypass first-pass metabolism. The protocol allows treatment with BHV-0223 to eligible adults who have Amyotrophic Lateral Sclerosis prior to it being available on the market.

Objective: Report on Zydis Riluzole 40mg ODT.

Methods: Of these 33 patients four are bulbar onset; 3 women and 1 man. 29 are limb-onset, 11 of which are women, 18 are men. 22 of the consented patients are currently taking drug. The mean of our initial ALSFRS-R is 33.5 and the mean of our Initial CNS-BFS is 44.77.

Results: Of 33 consented patients, 4 never started the BHV0223 22/29 patients who started the drug have been continuous with their dosing (75.8%); 3/29 patients had intermittent stop(s) but have continued with drug (10.7%), and 4/29 patients stopped drug completely (13.7%). To date 3 deaths of patients who started drug 2/25 on drug (8%) and 1/4 off drug (25%). Hypoaesthesia/numbness and tingling are possible side effects of taking the sublingual Riluzole. All of our patients have reported some sort of sensation, whether it be the hypoaesthesia or tingling separately, or together, after administration of sublingual Riluzole. It typically effects the tongue (posterior region), lips and/or entire mouth, as well as the area under the tongue, where the capillaries are trandporting the absorbed drug. We have patients who have symptoms anywhere from less than five minutes to almost one hour.

Discussion and conclusion: 7 of our consented patients (21.2%) who did take standard Riluzole tablets prior to beginning this clinical study expressed the ease in taking this drug more than the original tablet. The limitations of the regular tablet, including the inability of many patients to swallow a tablet formulated for oral administration, a negative food effect limiting bioavailability that forces a requirement for fasting, and hepatic toxicity related to dosing. Subjects adapted well to Zydis Riluzole 40mg oral disintegrating tablet. Early discontinuation was more common than late discontinuation.

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CMS-29 Real-world evidence of Radicava® (edaravone) for amyotrophic lateral sclerosis from a national infusion center database in the United States

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Keywords: clinical care, edaravone, care team

Background: Radicava® (edaravone) was approved by the United States (US) Food and Drug Administration for the treatment of amyotrophic lateral sclerosis (ALS) in May 2017 and became available to US health care providers in August 2017 (1). A pivotal, randomized, controlled clinical study conducted in Japan showed that edaravone slowed the rate of functional loss in ALS (2). Radicava® is administered by infusion at clinic sites, infusion centers, or at home. As 1 of only 2 therapies approved for the treatment of ALS in the US, and because the pivotal clinical studies for edaravone were conducted in Japan, there is interest in the real-world experience with Radicava®; however, there has been a paucity of real-world evidence (RWE) on the use of Radicava® in the US to date.

Objective: To provide RWE of Radicava® collected by a provider of home and alternative-site infusions.

Methods: A provider of home and alternative-site infusions and specialty pharmacy services in the US, Soleo Health (3), collected a database of RWE on the use of Radicava®. A retrospective analysis of this database was conducted, which included data from 123 ALS patients receiving Radicava® for at least 3 months. Analysis parameters included patient demographics, disease characteristics, ALS Function Rating Scale-Revised (ALSFRS-R) scores, quality-of-life and wellness information, and reports of adverse drug reactions. Usage patterns for Radicava® were also analyzed.

Results: The patients receiving Radicava® had a mean age of 59 years. The patients had been using Radicava® for an average of 193 days per patient. A complete analysis will be presented in the final poster.

Discussion and conclusion: This is the first report of RWE on the use of Radicava® in the US from data collected by an infusion and specialty pharmacy services provider. The data are expected to be helpful for clinicians who are considering using Radicava® therapy with their ALS patients.

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CMS-30 A preliminary analysis of the feasibility and efficacy of edavarone at a multidisciplinary ALS clinic

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Keywords: symptom management, clinical care

Background: Edavarone is an FDA approved, long-term infusion treatment demonstrated to slow disease progression in a Phase III clinical trial in individuals with ALS. Although edavarone was efficacious, the Phase III trial utilized highly restrictive inclusion criterion and the generalizability of results is currently unknown. Further, due to the high cost of treatment, insurance approval and drug administration represent barriers in the integration to clinical care.

Objective: To report the clinical feasibility and efficacy of edavarone treatment at a single multidisciplinary ALS clinic from August 2017-May 2019.

Methods: Thirty-seven individuals with ALS were clinically prescribed edavarone treatment. Demographics, treatment duration and infusion location were collected for all patients who initiated treatment. Routinely collected clinical endpoints including ALS-Functional Rating Scale scores, forced vital capacity and concomitant ALS-related treatments were aggregated pre and post-edavarone treatment using a clinical software platform (CareSense, MedTrak Inc). Descriptive statistics were utilized to compare changes in progression slope within individual patients across pre and post-edavarone treatment time points. Treatment was considered efficacious in those who demonstrated improved ALSFRS-R rate of change scores pre vs. post-edavarone.

Results: Of the 37 individuals with ALS prescribed edavarone, 59% ($n = 22$) initiated treatment. 64% ($n = 14$) of patients received home infusions and 36% ($n = 8$) received treatment at an outpatient infusion center. Primary reasons for not initiating treatment included: patient decision ($n = 3$), hospice ($n = 3$), insurance coverage ($n = 4$) and deceased prior to treatment approval ($n = 5$). Average pre- and post-treatment durations were 5 months (range: 2–11) and 10.45 months (range: 1–21), respectively. Overall, there was no change in disease progression rate pre and post-treatment (1.39 vs. 1.22 points per month). Of the 12 individuals with both pre and post- edavarone clinical data, 17% were considered responders ($n = 2$), with improved rates of ALSFRS-R change from 2 (pre) to 0.27 (post), and 0.7 (pre) to 0.2 (post) points per month. These individuals were also on riluzole. Fifty-eight percent of individuals demonstrated no benefit from treatment ($n = 7$) and 25% of individuals demonstrated hastening disease progression rates ($n = 3$). Individuals with mild-moderate disease (ALSFRS-R > 30, $n = 5$) did not respond to treatment (pre: 0.806 vs. post: 2 points per month).

Discussion and conclusion: Overall, edavarone was not an efficacious treatment in this descriptive analysis. In terms of disease progression and prescription of edavarone treatment, strict criterion were not utilized to identify eligible patients; however, individuals with milder, earlier onset disease did not respond better to treatment.

Acknowledgments: This study was sponsored by the Phil Smith Neuroscience Institute and Clinical Research Center.

CMS-31 Discussing personalised prognosis in amyotrophic lateral sclerosis: development of a communication guide

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Keywords: prognosis, communication, communication guide

Background: The recently developed personalised ENCALS survival prediction model for amyotrophic lateral sclerosis (ALS) makes it possible to reliably estimate life expectancy at diagnosis (1). Concerns were raised on how to discuss personalised prognosis in ALS without causing anxiety and destroying hope while meeting patients' needs (2). In other terminal diseases the topic is often avoided because of physician stress, lack of training and fear of causing distress and taking away hope (3). However, a majority of patients with ALS (66%) prefer a personalised estimate of their life expectancy (1).

Objective: To address these concerns and ensure a careful implementation, ALS Centre Netherlands tasked a multidisciplinary workgroup of neurologists, rehabilitation physicians and researchers to develop a communication guide to support neurologists and rehabilitation physicians in discussing the personalised prognosis with patients with ALS.

Methods: The workgroup inventoried important topics in discussing personalised prognosis: effect on patient's wellbeing; patient needs including needs of non-western patients; diverging information needs of patient and family; patients

with Frontotemporal Dementia (ALS-FTD). A systematic review was conducted to quantitatively and qualitatively synthesize evidence on effects of and needs for discussing prognosis in patients with a life-limiting disease. The workgroup together with an expert panel (patients, caregivers, ethicist and spiritual counsellor) developed recommendations based on evidence and expert opinion using consensus procedures.

Results: Evidence synthesis showed that discussing life expectancy had no negative effect on patients, if tailored to patient readiness. Themes regarding patient needs included honest communication, tailored information respecting cultural norms and values, hope-giving, family support and family-mediated communication. The workgroup drafted recommendations on using and interpreting the prediction model, communicating estimated life expectancy based on prognostic groups in a realistic manner while simultaneously conveying uncertainty of estimates and fostering hope, tailoring information to individual needs in a culturally sensitive way, and discussing prognosis in patients with ALS-FTD and in the case of diverging information needs. Recommendations were discussed with the expert panel and revised accommodating their comments.

Discussion and conclusion: Through multiple rounds of consensus procedures a communication guide was developed to support neurologists and rehabilitation physicians in discussing personalised prognosis with patients with ALS and their informal caregivers. The need for individual tailoring was emphasized and recommendations were also formulated on using and interpreting the online ENCALS survival model. Concluding, personalised prognosis can be discussed with patients with ALS if and when they desire and recommendations are provided on exploring and tailoring discussion to individual patients' needs. the guide can support physicians in tailoring discussion of personalised prognosis to individual patients' needs.

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CMS-32 A comprehensive approach to end of life discussions in amyotrophic lateral sclerosis

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Keywords: end of life, palliative care

Background: Amyotrophic Lateral Sclerosis (ALS) has a life span of 2–5 years after symptom onset. International consensus for optimizing patients with ALS (PALS) care includes utilization of interdisciplinary clinics. A part of this is a health care plan concerning end of life decisions.

Objective: Develop a coordinated effort between interdisciplinary clinic staff, home care companies, hospice providers, and ancillary staff, including palliative care physicians and chaplains, to assist PALS and caregivers in end of life decision making.

Methods: Multiple interventions were implemented to assist PALS and their caregivers in end of life health care decision making: 1. A simulation based training for health care staff in delivering the diagnosis of ALS was implemented. This training was followed by a debriefing and subsequent independent review of videos to help improve communication skills; 2. A dashboard was developed and utilized by the home care respiratory team to notify clinic staff if the vital capacity approached 30%, or ETCO2 of 45 or higher. When this occurred, the clinic staff would schedule immediate follow up clinic visit to have additional discussions of end of life planning; 3. A monthly support group coordinated by the nurse coordinator, social worker and respiratory therapist was developed for PALS and caregivers. This support group produced many open discussions about advanced care planning between the participants, moderators, and invited speakers.

Results: Patients and caregivers have benefited from improved communication, in home support, and monthly support groups. Total of 49 patients died since interventions started in 2017. There were 43 patients living in their homes prior to death. Six patients were residing in a nursing facility. Hospice involvement in home deaths occurred in 86% of PALS. One patient died in acute care hospital and two died in patient palliative care unit. Three patients were lost to follow up.

Discussion and conclusion: Improving communication skills in health care staff, implementing a home health dashboard, and developing a monthly support group have been well received. Additional changes to improve advanced planning have now been added upon feedback from PALS. There is now an ongoing clinical presence of a Palliative care physician available at team visits.

Acknowledgments: We would like to acknowledge the Harper's Hope Fund for ALS at the Medical College of Virginia Foundation which helped to financially support this work. We would like to acknowledge our patient and their families for their commitment to bringing awareness of the disease.

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