CMS-01: 10 years of riluzole use in a tertiary ALS clinic

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Live Poster Session A, December 9, 2020, 5:10 PM - 5:50 PM

Background:
Riluzole is a glutamate inhibitor approved for the treatment of ALS. There is scant data regarding factors associated with riluzole initiation and adherence.

Objectives:
Our goal was to describe the use of riluzole at the Penn State Hershey Medical Center (PSHMC) ALS clinic.

Methods:
We performed a retrospective medical record review of ALS patients seen at PSHMC from January 2007 to December 2016. Inclusion criteria were 1) Riluzole prescribed; 2) documented follow up until death, tracheostomy, or for at least 6 months following riluzole prescription. Demographic data and clinical history were extracted, including the dates of first and final neuromuscular clinics, King's College stage and ALS Functional Rating Scale-Revised (ALSFRS-R) at these visits, and date of placement of percutaneous endoscopic gastrostomy (PEG), tracheostomy, and death.

A timeline of riluzole use was established for each patient. Factors contributing to dose changes or discontinuations were recorded. Riluzole adherence was assessed using the proportion of days covered (PDC) calculated by the patient-reported length of riluzole use divided by total time from prescription to death/censor. Multivariable analysis was performed using Cox's proportional hazards regression to evaluate the association of demography and clinical course with adherence.

Results:
723 records were screened, with 437 (264 men, 173 women) meeting criteria for inclusion. The median age at onset was 61.6 years. The median ALSFRS-R at first evaluation was 39, which declined to 18 at the final evaluation. The median tracheostomy-free survival after the date of disease onset was 1068 days. 410/437 patients (93.8%) started riluzole. 39 (9.5%) changed dose a total of 66 times, most commonly for abnormal liver function tests (n=16) or gastrointestinal side effects (n=6). 115 patients (28.0%) discontinued riluzole a total of 127 times, most frequently citing gastrointestinal symptoms (n=20), cost (n=18), patient preference (n=17), hepatotoxicity (n=15), and advanced disease/hospice (n=12).

The median length of riluzole use was 435 days (range 0-3773). The mean PDC for the group was 62.8 ±27.2%, with 266 individuals (64.9%) demonstrating a PDC greater than or equal to 60%. Those younger at the time of prescription, with higher ALSFRS-R and slower rate of decline were more likely to have a larger PDC. No trends in patient demographics, riluzole use, and tracheostomy-free survival were found over time.

Discussion:
We found a high rate of riluzole initiation and adherence, with a large majority of prescribed patients initiating riluzole and nearly 65% demonstrating a PDC > 60%. The most common reasons for dose modification were related to adverse effects, though social, economic, and patient related factors were also common. The characteristics of riluzole prescription and use have remained relatively unchanged in a single tertiary ALS center over the last ten years.

CMS-02: A cost effectiveness framework for amyotrophic lateral sclerosis, applied to riluzole.

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Background:
Approximately 5,000 patients develop amyotrophic lateral sclerosis (ALS) annually in the US. ALS is an expensive disease, with annual per-patient and national costs exceeding $70,000 and $1 billion respectively. A increased interest in health economic evaluations of treatments of ALS is anticipated with active development of new treatments.

Objectives:
(1) Reexamine cost-effectiveness of riluzole in the treatment of ALS in light of recent advances in disease staging and understanding of stage-specific drug effect.
(2) Thereby propose a consistent framework for future health economic evaluations of ALS treatments.

Methods:
ALS was staged by the Fine'til 9 (FT9) Staging method. Stage-specific health utilities (EQ-5D, US valuation) were estimated from an institutional cohort, whereas literature informed costs and transition probabilities. Costs at 2018 prices were disaggregated into recurring costs (RCs) and "one-off" transition/"tollgate" costs (TCs). Five- and 10-year horizons starting in stage 1 disease were examined from healthcare sector and societal perspectives using Markov models to evaluate riluzole use, at a threshold of $100,000/QALY. Probabilistic and deterministic sensitivity analyses were conducted.

Results:
Mean EQ-5D utilities for stages 0-4 were 0.79, 0.74, 0.63, 0.54, and 0.46, respectively. From the healthcare sector perspective at the 5-year horizon, riluzole use contributed to 0.182 QALY gained at the cost difference of $12,348 ($5,403 riluzole cost, $8,870 RC and -$1,925 TC differences), translating to an incremental cost effectiveness ratio (ICER) of $67,658/QALY. Transition probability variation contributed considerably to ICER uncertainty (-30.2% to + 90.0%). ICER was sensitive to drug price and RCs, whereas higher TCs modestly reduced ICER due to delayed tollgates.

Conclusions:
This study provides a replicable framework for health economic studies of ALS treatments using FT9 staging that can be readily applied to retrospective clinical trial data. Prospective stage-specific and disaggregated cost measurement, however, is warranted for accurate future cost-effectiveness analyses, because appropriate separation of TCs from RCs substantially mitigates the high burden of background cost of care on the ICER.

CMS-03: A sticky problem: The management of thick oro-pharyngeal secretions in amyotrophic lateral sclerosis

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Background:
Thick, sticky, oro-pharyngeal secretions can be a distressing problem for patients with amyotrophic lateral sclerosis (ALS). There is no evidence base to guide treatment which is often managed mostly by clinician experience. Options for treatment for thick secretions traditionally include conservative management, carbocisteine and beta blocker. Treatment of sialorrhoea can cause thickened secretions therefore it is essential to consider the balance of thick and thin secretions in an individual when considering treatment options.

Objective:
To describe the response of two patients to nebulised therapies for thick oro-pharyngeal secretions.

Methods:
Hypertonic Saline (HTS) and N- acetyl cysteine (NAC) nebulisation are regularly used treatments for secretion management in cystic fibrosis. We report a case study of two MND patients prescribed HTS for oro-pharyngeal secretions. The treatment ladder consisted of an initial trial of 3% hypertonic saline, increased to 7% HTS and finally adding NAC as a final step if necessary (all delivered via nebuliser). Participants were assessed at 3 monthly intervals using their CSS-MND score, a patient reported outcome measure for which a higher score indicates more severe saliva symptoms (max score 30) and a global change questionnaire (GCQ) which assessed the change in impact of their saliva problems compared to three months earlier. As 7% HTS and NAC nebulisation can cause bronchospasm, patients had their 1st test dose delivered in conjunction with Salbutamol in a clinic setting.

Case 1:
A 44-year-old gentleman with ALS developed thick secretions due to poor swallowing and ineffective cough. He followed conservative management approach and tried carbocisteine at maximum dose without much benefit. Nebulisation with 3% HTS offered no improvement. 7% HTS was prescribed a month later which yielded significant improvement in both outcome measures (CSS – MND, GCQ). The effect of HTS nebulisation plateaued after ~18 months, NAC nebulisation was offered. This was not deemed to be more effective and patient preferred to return to 7% HTS.

Case 2:
This 50-year-old gentleman with ALS initially presented with excessive thin secretion complicated by postnasal drip causing major difficulties with NIV usage. To manage this, he was tried on hyoscine, propantheline and atropine with some improvement but complicated by thick secretions. He tolerated 3% HTS well with slight improvement in the CSS – MND score. A trial with 7% HTS did not yield better outcome than 3% HTS, no reduction was seen on amended CSS-MND score, consistent with a lack of improvement reported by the patient in GCQ. This patient was not trialled on NAC.

Conclusion:
Managing both thick and thin secretion effectively in MND patients remains a challenging issue. We present a treatment approach using nebulised HTS and NAC. Further research is needed to develop the evidence base in this area.

CMS-04: ALS Focus–A new survey program for people with ALS and caregivers: Results from Survey 1 on insurance needs and financial burdens

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Background:
ALS Focus is a patient- and caregiver-led survey program to understand the experiences, needs, and preferences of people living with ALS and caregivers in the United States (www.als.org/ALS-Focus).

The U.S. Food and Drug Administration (FDA) calls for rigorous input from patients and caregivers to inform clinical outcome assessments in drug development. This research domain, called patient focused drug development (PFDD), identifies and validates clinical outcomes by asking patients and caregivers directly what matters most to them (FDA, 2018). In the ALS space, scientifically rigorous efforts to collect this experience and preference data are in their infancy (Brizzi et al., 2020). With direct insight from a patient and caregiver advisory committee (PCAC), ALS Focus survey research is designed to address this gap in PFDD specific to ALS. De-identified Focus data are open-access and free to use. NeuroSTAmPs are generated for all participants to connect Focus data to other studies.

The first Focus survey measured insurance needs and financial burdens that people with ALS and caregivers experience when confronting the disease.

Methods:
The ALS Association develops ALS Focus surveys with input from people with ALS, caregivers, and industry and academic experts. Surveys are issued throughout the year and are available online through an account-based portal at the Massachusetts General Hospital's (MGH) Neurological Clinical Research Institute (NCRI).

The first survey took place from February to April 2020. People with ALS, current caregivers, and past caregivers who were 18 years old or older and lived in the United States were eligible to take the survey after consenting. Participants also completed demographics and disease history questions.

Results:
The final sample for the survey on insurance needs and financial burdens included 204 people living with ALS, 116 current caregivers, and 120 past caregivers (N = 440). Nearly a quarter of participants reported working beyond when they had planned after an ALS diagnosis, 42% of whom did so to maintain health insurance. Of the participants who reported losing health insurance after an ALS diagnosis (10%), 67% lost insurance when they had to stop working. Responses showed home modifications were among the most needed, highest cost, and most burdensome expenses that participants experienced. Finally, nearly 40% participants with ALS reported high stress in reaction to covering the costs for medical treatments and services, understanding their health insurance coverage, and managing medical billing paperwork. This presentation will describe remaining survey results, and implications for policy, care, and future PFDD research.

References:
Brizzi, K. T., et al. (2020). Understanding the needs of people with ALS: A national survey of patients and caregivers. Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration, 21(5-6), 355-363. https://doi.org/10.1080/21678421.2020.1760889

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CMS-05: An interdisciplinary approach to mindfulness, in the time of CoVid-19, as a quality of life improvement factor for health care professionals, people with ALS, and their primary caregivers

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ALS is a disease of losses and adjustments. People with ALS (PWALS) face higher than normal anticipatory grief specific to lost abilities, social interaction and life. The need to readjust, grieve and reinvent oneself in the face of ongoing change is a constant.

The idea that the world is in a state of constant change, that is, nothing endures in a static state, is the epistemological background of mindfulness. Mindfulness is being aware that everything can be perceived from different perspectives. This habit of perception can be learned, and several decades of study indicate that such learning may increase the ability to navigate grief and benefit quality of life (QoL). Recent studies using a Langerian mindfulness framework (LMF), suggest that greater mindfulness in PWALS and their primary caregivers (PC) correlates with a higher QoL.

Interdisciplinary care has been shown to be the best model of care for ALS, with effective clinical care being built over time, through in person clinical visits and telephone contacts. However, the CoVid-19 pandemic has altered both care delivery and stressors that healthcare professionals (HCP) are facing.

Many HCP are experiencing levels of uncertainty, loss and rapid change, that parallel those regularly experienced by PWALS/PC. Studies have identified several areas of stress for HCP, including risks of infection, lack of personal protective equipment, increased workloads, insufficient personnel, redeployment, isolation, separation from families and burnout. There are concerns that these pressures may have both short- and long-term consequences on mental health and QoL.

The Montreal Neurological Institute and Hospital (MNI/H) ALS program seeks to contribute to QoL in HCPs and PWALS/PC by 1) training HCPs on the LMF, 2) adapting the LMF for the MNI/H ALS program, and 3) conducting an action research study in PWALS/PC to investigate whether a mindfulness protocol increases and maintains mindfulness, and in turn influences QoL.

Training HCPs on mindfulness, to later deliver it as a clinical practice, may reduce immediate and long-term stress, maintain QOL, and improve empathetic understanding of PWALS/PC stress and grief. In other words, increase HCP understanding of the mindfulness needs regularly faced by PWALS/PC. Furthermore, having HCPs undertake mindfulness training could accrue broader applicability in the transfer of it as a clinical practice, and skillset.

The interdisciplinary nature of the MNI/H ALS program presents a unique occasion to refine and test a mindfulness-based intervention, which can serve as a model for multiple settings within the ALS community. Given that there is no cure for ALS and that available pharmacological interventions only target slowing of disease progression and provide a modest increase in life span, any applicable skill set that facilitates maintenance and/or improvement of QoL may indicate a best practice approach.

CMS-06: ALS Patient and Caregiver Attitudes Toward Physician-Assisted Death in California

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Background:
In the United States, Oregon became the first state to legalize physician-assisted death (PAD) in 1997. In the United States, 8 states and the District of Columbia now have laws legalizing PAD, including California, which began allowing PAD through the End of Life Option Act (EOLOA) in 2016 (1). Amyotrophic Lateral Sclerosis (ALS) specifically is a disproportionally common diagnosis for patients who have used the EOLOA in California, despite being a rare disease (2). Because PAD has recently become legal for 70 million people in United States, including 40 million Californians, we perceived a need to assess the attitudes of ALS patients and their caregivers toward PAD, gaining insights into reasons why they would consider using PAD.

Objectives: To quantify how many patients with amyotrophic lateral sclerosis (ALS) and their caregivers have considered physician aid in dying (PAD) through the End of Life Option Act (EOLOA) in California and assess reasons to consider using this act.

Methods:
We performed a cross-sectional study without external control at one ALS Center. We surveyed patients with ALS and their caregivers on their views about the EOLOA. Data on disease characteristics, demographics, quality of life, and depression were also collected, via validated questionnaires as applicable. Descriptive statistics were used to analyze the data, along with the Student's t-test.

Results:
A small majority of ALS patients surveyed would consider using the EOLOA (53%, 16/30). Similar rates of caregivers thought their loved one would consider using the EOLOA (40%, 11/27). Patients most commonly described having intolerable symptoms, being a burden on their loved ones, and losing independence as reasons to consider using the EOLOA. Many patients shared that "their life has purpose" and "they are making the most of their lives" as to why they are not considering the EOLOA. Considering the EOLOA was not related to disease severity (forced vital capacity, p=0.84 or ALSFRS-R score, p=0.58) or depression (p=0.26).

Conclusions:
Pursuing PAD is a personal decision for each individual patient. This study shows that considering PAD is relatively common in ALS patients, independent of disease severity or presence of depression.

References:
1. End of Life Option Act, California, 2015.
2. Nguyen et al. Jama IM 2018. 178:417–421.

Acknowledgements:
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CMS-07: Canadian Best Practice Recommendations for the Management of ALS

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Objective:
Recognizing the need to establish a national standard for management of patients living with ALS in Canada, Canadian ALS clinicians sought to develop best practice recommendations. The guidelines update evidence provided in previously published guidelines and address issues unique to Canada. The recommendations are supported by evidence or by Canadian expert consensus where evidence is unavailable.

Background:
Health care in Canada is mandated by the federal government but delivery is the responsibility of the provinces and territories. Although, several ALS clinical practice guidelines have been published (AAN ALS Practice Parameters (2009), EFNS ALS guidelines (2011), and NICE Motor Neurone Disease guidelines (2016). No published Canadian ALS clinical guidelines set a national standard for care of patients with ALS.

Methods:
The guideline working group consisted of eleven Canadian ALS clinicians with geographic representation from across the country. Clinical questions of interest were obtained by surveying clinicians and staff at all the Canadian ALS clinics, and then further refined by the working group. Literature searches on the questions were conducted by a consulting firm (Centre for Effective Practice) with guideline development experience. Medline, EMBASE, and CINHAL databases were searched. Retrieved abstracts were screened for inclusion criteria, and selected publications were reviewed for relevance and data quality. Guideline statements for each clinical question were developed on an iterative basis until consensus was obtained. Finally, each guideline statement was assigned an evidence rating, which included the option of expert consensus. Public feedback on the draft recommendations was elicited, discussed, and revisions were discussed by the writing group.

Results:
Canadian ALS Best practice recommendations include guidance on: 1) delivery of an ALS diagnosis, 2) disease related treatment, 3) multidisciplinary care, 4) respiratory management, 5) nutrition management, 6) DVT risk, 7) medication alignment, 8) symptom management, 9) dysarthria management, 10) exercise in ALS, 11) cognition, 12) caregiver burden and support, 13) palliative care, and 14) medical assistance in dying. The respiratory statements were harmonized with the Canadian Thoracic Society recently published guidelines for home mechanical ventilation for patients with ALS. Unique aspects of these guidelines are statements concerning edaravone use, recommendations for timeliness of interventions, early interventions for respiratory insufficiency, thromboembolism screening, regular review of medications and discontinuation of non-essential medications, exercise prescriptions for patients with ALS, and approach to medical assistance in dying requests.

Conclusions:
These recommendations will serve to better support the clinical care and management of Canadian patients living with ALS by updating existing guidelines and providing consensus recommendations where evidence is insufficient.

Acknowledgements:
The authors are grateful for the ongoing support from ALS Canada and Canadian ALS Clinical Research Network (CALS) for the development of these guidelines.

CMS-08: Caregiving and Sleep Disruption: An Assessment of Young Carers in ALS/MND

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Background:
Sleep-wake disruption is common among 'informal family caregivers', and is more prevalent than in the general adult population. However, these disruptions remain largely unknown in a vulnerable and isolated subset of caregivers – children and youth under the age of 18. These 'young carers' provide care across the disease spectrum and care need, including amyotrophic lateral sclerosis (ALS).1 Caregiving impacts youth school attendance and performance, and managing their own mental well-being, yet the health effect effects, including sleep, are largely unknown2.

Objectives:
(1) Assess the feasibility of measuring sleep quality in caregivers and non-caregivers, and (2) Identify differences in sleep health related quality of life between young caregivers and non-caregivers over a 5 day, 24-hour sleep-wake cycle

Methods:
Quasi-experimental design, with age and gender matched treatment and control groups. Caregiving youth "treatment" (n=7) recruited via the ALS Multidisciplinary clinic and local Chapter of the ALS Association. Non-caregiving youth (n=13) were recruited word of mouth via the research team. Participants wore an GENE-active actigraphy device, and kept a daily journal of all activities for 5 consecutive 24-hour periods. Study measures included demographics, caregiving tasks, Pittsburgh Sleep Quality Index, and sleep-wake measures: 1) Self-reported bedtimes and awake times, 2) total sleep time and the number of minutes awake after sleep onset. Rest-activity rhythm (RAR) parameters modelled from minute-to-minute actigraphy count data by a five-parameter cosine model with an anti-logistic function.

Results:
All participants wore the watch and completed their journal for the full 5 days. Caregivers participated in an average of 7 caregiving tasks, including feeding, bathing, and transferring. caregivers have shorter sleep duration (t = 51.19 (11.99), latency (t = 52.42 (10.74), efficiency (t = 55.49 (14.00) , and overall sleep quality (t = 51.32 (12.26). Caregivers had lower total sleep time (CG = 6.75 ± 1.47, NCG = 7.08 ± 1.36) and sleep efficiency than non-caregivers (.80 ± .23)).

Discussion:
This data is the first to track sleep in young caregivers, and may serve as a baseline to assess future cascading health impacts of care associated with disruptive sleep. Understanding how care disrupts sleep is critical towards lessening the potential for long-term impacts of caregiving on youth health and well-being, including obesity, anxiety and depression. Results have clear practice implications for health and social care providers and the creation of targeted health and social interventions for young caregivers.

References:
1. Kavanaugh MS, Cho C, Howard M., et al Neurology; 2020
2. Kavanaugh MS, Stamaopoulos V, Cohen D et al Adolescent Research Review 2015; 1(1), 29-49.

Acknowledgements:
We would like to thank the families and youth who gave of their time for this project. Funding for this study was provided by the Clinical and Translational Science Institute of Southeastern Wisconsin

CMS-09: Does the use of a patient passport improve patient care in MND? A patient's perspective

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Background:
Care provision for Motor Neurone Disease (MND) patients require a Multidisciplinary Team (MDT) approach. Healthcare Professionals (HCPs) and MND patients often find it challenging to communicate effectively, particularly during the current COVID19 era. Communication is particularly difficult and discontinuous in patients with rapidly changing needs. Patient Passports (PP) can be used as a tool to improve communication and thus patient experience. It is designed to provide brief but vital information about the patient. We studied the place of PP in MND care. During COVID-19, there has been a disruption to the regular care provided to MND patients. High level communication of change in care needs to the HCP is important.

Aim:
To explore the views of MND patients and staff involved in MND care regarding the use of PP.

Methods:
MND patients (n=30) and staff involved in MND care were consulted using semi-structured interviews, questionnaire and focus group discussions.

Results:
Overall, themes emerged overwhelmingly favoured the use of a MND passport. These include improvement in communication (47%), allowing for greater coordination of care (31%) and standardising healthcare provision across different regions (22%). The emergency care providers and community HCP's felt the PP was paramount to communicate with the parent team. However, concern was raised for the need to tailor it to the patient.

Conclusion:
The PP was recognised as a distinct need. An online, updatable passport could be a tool set in place to facilitate coordinated and timely patient care. However, to validate the above findings, wider MND patient/carer and HCP survey is necessary.

CMS-10: Effectiveness of eye-tracking systems for communication with ALS patients

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Aims:
Late-stage ALS patients frequently suffer from a progressive communication impairment, which in most cases, coexists with a preserved cognitive functioning. In the locked-in state (LIS), eye-tracker systems (ETS) restore verbal communication and computer-control for several applications such as work, entertainment, social network participation, and home automation. However, only a small part of LIS patients successfully use at home these sophisticated devices, and little is known on factors predicting the efficacy of ETS control in this group of patients (1).
In this study, we report the performance parameters obtained by a cohort of non-LIS ALS patients in the first spelling session with an ETS in comparison to healthy control.

Subjects and methods:
We enrolled 61 ALS patients referred by the ALS Tertiary Clinical Research Center of the University of Palermo (Age 61,9±10.32 years; Education 10.67 ± 4.7 years) and 14 healthy relatives of them (Age 48.35± 18.40, p= 0.38; Education 10.45 ±4.39, p= 0.5). Patients were disabled but not in an advanced stage of the disease (ALSFRS-R 32,68 ± 9.38). All subjects were asked to write a single 13-letter word (DELICATAMENTE) using an ETS. Accuracy (number of correct letters/total letters %), Execution time, and Letters per Minute (LPM) were recorded and related to ALS patients demographic (age, education), clinical (disease's duration, ALSFRS-R), and neuropsychological variables (Frontal Systems Behavior Scale [FrSBe], Edinburgh Cognitive and Behavioral ALS Screen [ECAS] and Beck Depression Inventory [BDI]).

Results:
In spite of being at their first ETS trial, both ALS patients and HC showed high precision spelling, with a mean Accuracy of 94.1±11 % in ALS patients and 92.2±10.9 in the HC group (p= 0.8). Similarly, Execution time was short and not different between ALS patients and controls (70±15.4 sec in ALS vs. e 72 ±18 sec. in HC, p=0.6), as well as the LMP (10.9±3.3 in ALS vs. 10.7±3.8 in HC, p= 0.3). In the ALS group, older age, functional impairment measured by ALSFRS-R, and cognitive/behavioural changes detected by ECAS were related to a longer Execution time. The other performance parameters were not affected by demographic, clinical, or neuropsychological variables.

Conclusions:
Most ALS patients at the intermediate stage of the disease are able to communicate successfully with an ETS, without needing long training. Older age, global functional impairment, and cognitive/behavioural changes are associated with longer Execution time. Consequently, the spelling parameters should be adapted to the patient's characteristics. It is not clear whether cognitive variables are the main responsible for the relatively small number of LIS ALS using regularly ETS. Future longitudinal analyses will explore the ETS performances over time up to the end-stage disease.

References:
1. Spataro R et al. Acta Neurol Scand 2014; 130:40-45

CMS-11: Genetic testing for ALS in Canada – An assessment of current practices

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Background:
Several therapies are currently in development for ALS with a genetic etiology. While genetic testing should be available for patients who have a clear family history of ALS (fALS), testing of seemingly sporadic cases (sALS) is not widespread. With the growing role of genetics and potential targeted therapies for ALS, it will be crucial to ensure that patients with a monogenetic etiology are identified in a timely manner.

Hypothesis:
Access to genetic testing, and genetic testing practices, are inconsistent across Canada, in a federally funded, provincially regulated health care system. This leads to the possibility that patients who could potentially benefit from emerging therapies are not being identified.

Methods:
ALS clinicians from both Canadian ALS Research Network (CALS, n=20) and non-CALS (n=4) clinics across Canada were contacted to assess their practices with regards to genetic testing. Clinicians completed an online survey, which was followed up with a semi-structured phone interview, tailored to their practices, clinic, and province of practice.

Results:
Not all clinics in Canada are routinely ordering genetic testing for fALS cases. One third of clinics are routinely ordering genetic testing for seemingly sALS cases. For clinics not routinely ordering testing for sALS cases, the most often cited reason is that there is a lack of implication for a treatment plan. Predictive testing practices are highly variable. The average wait time for an ALS patient to see a genetic counsellor in Canada is 9.5 months (range 0-36 months). Families with known ALS mutations are not prospectively being followed.

Discussion & Future Directions:
Genetic testing practices, and access to testing, are inconsistent across Canadian ALS clinics. There may be patients with a monogenetic etiology to their ALS who are not being identified. Barriers to genetic testing vary from province to province, and include provincial ministry of health approvals, institutional approvals, and access to genetic counselling. ALS clinicians have a strong interest in both early and late-stage clinical trials for the genetic forms of ALS, and a desire for consensus guidelines to be developed.

The goal of this work is to implement a standardized approach to genetic testing for ALS patients across Canada. A quality improvement plan is being designed, which will implement a mainstreamed approach to genetic testing in ALS, in ALS clinics in Canada, in order to determine feasibility of widespread genetic testing in a federally funded health care system.

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Acknowledgements:
We would like to thank the clinicians who participated in both survey completion and phone interviews for their time and enthusiasm for this initiative. Funding for this work has been provided by Biogen, through a Sponsored Research Agreement.

CMS-12: Telehealth approach for amyotrophic lateral sclerosis patients: lesson from COVID-19 pandemic

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Live Poster Session A, December 9, 2020, 5:10 PM - 5:50 PM

Background and objective:
Specialized multidisciplinary ALS care has been shown to extend survival and improve patients and caregiver's quality of life. During COVID-19 pandemic, the management of patients suddenly changed and telemedicine has been proven to be useful for monitoring patients. Here we report the experience with Telemedicine of a Tertiary ALS Center from an Italian geographical area with high infectious risk during COVID-19 pandemic.

Methods:
We focused the project on the following areas of intervention, based on the most common set of problems in ALS patients: 1) Global Function and mobility; 2) Respiratory Function; 3) Nutrition; 4) Communication, Psychology and Cognition.
During COVID-19 pandemic 19 patients were evaluated in telemedicine by a multidisciplinary team including: neurologist (clinical evaluation, intercurrent events, and drug prescriptions); dietician (diet and weight monitoring); psychologist (psychological assessment and support); physiotherapist (physiotherapy treatment and device prescription). All patients were in the first year of disease.
Telemedicine was performed using the online platform "IoMT Connected Care Platform (Ticuro Reply)".

Results:
Firstly, all patients reported a positive perception of talking face-to-face with healthcare professionals and were satisfied on how the team understood their problems. During a video televisits, there was a change in patient's medication regimen in 11/19; 2/19 required in-hospital pneumological evaluation and started Non-Invasive Ventilation; 9/16 patients required devices prescription. The mean monthly decline of ALSFRS-R before televisit was 0.88 (SD 1.17), and during televisit of 0.49 (SD 0.75). Body weight and daily caloric content remain stable. Reduction in HADS scores and stability in ALSAQ-40 were observed. None of the patients developed COVID-19 symptoms during the study period.

Discussion:
With this pilot trial, we defined the feasibility and efficacy of technology support in monitoring patients with chronic diseases, even in a tragic period as the COVID-19 pandemic. We positively reproduced the standard multidisciplinary approach currently used, stabilizing the functional and metabolic status and improving the psychological one.
Based on these positive results, we started a new trial with the aim to include telemedicine in a personalized multidisciplinary program according to the patient needs, which is adapted to the patient both in terms of televisit frequency and healthcare providers involved. In this part, we are enrolling patients in different stage of disease, with various needs, to receive additional tele/technology supports from a targeted specialist, comparing them with a control group patients - matched for age, sex and clinical stage – that are followed with the traditional in-person multidisciplinary care.

CMS-13: When months matter; the projected impact of the COVID-19 pandemic on the UK diagnostic pathway of MND

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Live Poster Session A, December 9, 2020, 5:10 PM - 5:50 PM

Background:
The MND diagnostic pathway is slow, taking on average 10-16 months from symptom onset (1). Early diagnosis is important to access supportive measures and advanced care planning with the aim of maximising quality of life.

The COVID-19 pandemic has caused significant delays in NHS pathways; many GP appointments occur via video with subsequent delays on secondary care appointments and investigations. Given the rapid progression of MND, patients may be disproportionately affected resulting in late stage presentations impacting local services.

We use Monte Carlo simulation to model the pre-COVID-19 diagnostic pathway. We then introduce plausible COVID-19 delays, exploring changes to the pathway, focusing on different MND phenotypes to explore whether delays result in more late-stage presentations. We adopt a predictive approach to allow primary and secondary care to adjust proactively.

Methods:
The diagnostic pathway was modelled using gamma distributions of time taken: 1) from symptom onset to presentation to GP, 2) for specialist referral, and 3) for diagnosis to be reached after neurology appointment. We incorporated a branch when the GP consultation did not result in a referral. An acute presentation triggered when diagnostic pathway time was within 30 days projected survival (based on phenotype). The total time-to-diagnosis was calculated over 105 iterations. The pre-COVID-19 model was estimated using data from the literature and Improving MND Care Survey 2019. We estimated delays due to COVID-19 using published statistics where available.

Results:
Pre-COVID-19 model is in keeping with published data (median time to diagnosis 15.5 months). Two COVID-19 models were used: primary care service times only increased by 50% and if all services are impacted. The model suggests a diagnostic delay of 9.3 months and 16.7 months. Emergency presentations were increased by: ALS, 24% and 39%, bulbar-onset, 25% and 41%; limb-onset, 5% and 18%.

Discussion:
The model suggests the COVID-19 pandemic will result in later diagnosis and more acute presentations for people with MND. Primary care and non neurology secondary care teams should consider late-stage presentations of MND when assessing patients. There are a number of possible solutions; the MND Red Flags GP checklist tool could be updated; development of rapid assessment clinics for suspected plwMND. Late-stage presentations may require rapid escalation to appropriate multidisciplinary and palliative care. Proactive recognition of acute and late-stage disease with altered service provision will optimise care for plwMND during the pandemic.

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1. Richards D et al. Time to diagnosis and factors affecting diagnostic delay in ALS. Journal of the Neurological Sciences. 2020.

CMS-14: Impact of COVID-19 pandemic lockdown in a Catalonian cohort of ALS patients

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

The SARS-CoV2 pandemic has produced a series of changes in the health system that has affected, most notably those patients with chronic diseases such as ALS.

To know the impact of COVID-19 lockdown in the care of patients with ALS in Catalonia, we have conducted a survey for patients and their caregivers, also to the interactions with the healthcare systems during this time, and acquire a better knowledge of their needs.

Of 57 patients, most of the responders were in a high dependency situation (49.12% in 4b King's stage). None of the patients and their co-habitant (familiars and caregivers) had COVID-19.
As important data, 21.43% had to change their usual care provider, 29.82% were unable to contact with their medical professionals and 68.42% did not have personal protective
equipment for their familiars or caregivers.

Knowing these data will help us make better preventive decisions which will result in better care for the patient. This knowledge is useful for proper planning in the event that similar situations arise again, as well as highlighting the degree of vulnerability and the need of increase efforts to enhance telemedicine in ALS patients.

CMS-15: Individual Quality of Life among spousal ALS patient-caregiver dyads

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

Background:
Quality of life is a basic goal of health and social care. It is important to recognize the factors that contribute to quality of life for individuals to better understand the lived experiences of this illness within a care management environment.

Objective:
To explore individual quality of life (IQoL) of people with ALS and their informal caregivers over the illness trajectory.

Methods:
In three semi-structured home interviews, quantitative and qualitative data were collected on quality of life (SEIQoI-DW) from 28 spouse patient-caregiver dyads.

Results:
Individual Quality of Life was high for both patients and caregivers with some fluctuation over time. IQoL was higher among patients than for their care partners at each interview. Psychological distress was higher among caregivers, and burden consistently increased. Family, hobbies and health were defined as contributors to quality of life most often. The importance of health declined relative to other life areas over time. Friends and finances became less important for patients, and both factors were assigned greater importance by caregivers.

Conclusion:
Experiences of the person with ALS and his/her informal caregiver represent an important source of information on the lived experience of the condition. The findings point to the importance of exploring and monitoring quality of life at an individual level, and the self-defined contributory factors relevant to the individual within his/her context. The contributing factors may change in form and content along the disease trajectory. IQoL used as an integrated outcome measure can facilitate conversations between health care providers, patients and families, and to inform interventions and care management.

Acknowledgements:
Research Participants; funding from the Irish Health Research Board.

CMS-16: Life 'on high alert': how do people with a family history of motor neurone disease make sense of genetic risk? Insights from an online forum.

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

Background:
Up to 10% of people with MND have an inherited form of the disease, knowledge that has far-reaching implications for other family members. In such cases, family members may not only face specific issues around their relative's diagnosis and care, but living with genetic risk. Few studies have explored experiences of people affected by inherited forms of MND, in particular using extant forum data to gain a naturalistic insight into the attitudes and concerns of such individuals and the interactions between them.

Objectives:
To explore how people using the MND Association Forum make sense of, and negotiate, genetic risk. In particular, this paper looks at how individuals construct risk through tracing their family history of the disease, at times interpreting this information to quantify their own risk; it draws out the different ways people living with the threat of MND express uncertainty over their future; and it seeks to understand how choices around pre-symptomatic genetic testing and reproductive decision-making play out on the forum.

Methods:
A thematic analysis of posts shared between 2010 and 2019 on the MND Association Forum, an online space where people affected by MND can seek information and support from others in a similar situation. Posts around inherited MND were identified using key word searching, and 37 threads containing 332 posts were included in this analysis. Posts were analysed using an indictive approach.

Results:
This paper highlights the experience of living at risk of inherited MND as crosscut by multiple uncertainties- around scientific developments and the future of the disease; their own genetic risk, which is often hard to quantify; and making decisions around genetic testing and reproductive options, amidst a complex biomedical and ethical landscape. In the context of this uncertainty, the forum is a site where experiential knowledge is widely foregrounded in the information and advice shared between users, becoming intertwined with (interpretations of) biomedical and genetic information. This paper pays attention to the forum as an interactional space where uncertainties are negotiated and risk is made sense of between individuals with a family history of MND, and those affected by 'sporadic' forms of the disease.

Discussion and conclusion:
This study attests to the value of online forums as repositories of rich data which can help us understand patient and family experiences. It highlights some of the concerns and challenges faced by people living with a family history of inherited MND.

Acknowledgements:
We wish to thank the MND Association for allowing us to use the publicly available information on the MND Association Forum for this research, and for their overall support of the study. Jade Howard is funded by a doctoral studentship provided by the Institute of Applied Health Sciences, University of Aberdeen.

CMS-17: Managing Community Motor Neuron Disease patients during the Covid 19 pandemic- practice changes

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

Background:
Health care provision throughout Covid-19 has been a challenge for patients and clinicians (1). Requiring rapid practice changes to clinically manage and support patients, (1, 2). Including assimilating new information whilst delivering gold-standard care and managing patient's uncertainty (1).

Objectives:
To discuss from the perspective of a community-based Specialist Palliative Care (SPC) service with an integral role in a city wide MND Multi-disciplinary team (MDT), how Covid-19 altered the clinical management and support of MND patients. Reflecting on these challenges, we highlight key themes that may help other providers shape their services for any further waves of mass infection.

Context:
The Coventry SPC Team is a multi-professional team. Forming a key part of the city's wider MND MDT incorporating professionals across hospital, hospice and community, managing patients with a confirmed MND diagnosis under a Coventry GP.

Method:
From March – Sept 2020 the service managed 19 patients; including 5 newly diagnosed and 3 existing patients deceased during the pandemic.
3 cases were selected to demonstrate the challenges of clinical management and patient support.

Key practice changes:-
Patient support during the specified period involved creative solutions to equipment and exercise provision and assessments. Introduction of virtual consultations (attend anywhere) whilst recognising clinical provision required some face to face (3)and increased use of telephone support for additional psychological support (2).
Clinical management involved practice changes with increased focus on contagion and risk management – PPE, Enhanced PPE (1, 3), Alerts on community/ nursing system re patient's vulnerable status. Being a visible contact point to reassure 'NHS' services remain accessible
Also aiding patients with signposting/ links: - re changing access to services e.g. GP, hospice, hospital. Advice re government shielding guidelines and impact on shopping, family and managing isolation (2).

Discussion:
Covid -19 has been a unique challenge for staff and patients. Maintaining regular contact for support and guidance has been essential to support patient's wellbeing through Covid (1). Moving forward, in terms of patient management, whilst informal mechanisms can capture the patient's voice, more formal mechanisms should be put in place such as questionnaires or online interviews, to establish which changes have been essential and beneficial, and which detrimental (3). Furthermore, to capture patterns a review of case reports could be carried out. Together helping drive appropriate ongoing service change and future provision for MND patients during this unique window of time.

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CMS-18: Multinational Public Policy in ALS/MND

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

Background:
There is a clear need for public policy and research focused on the needs of patients with ALS/MND and their families. There is also a need for comparative information regarding public policy and its influence on ALS/MND clinical care and research.

Introduction:
ALS/MND public policy is expected to vary widely among countries across the globe. Key determinants of public policy include economic, cultural and religious factors. Significant differences should occur when comparing WHO-designated high-, low and middle-income countries.

Methods:
More than fifty ALS/MND experts from 21 countries around the world provided key information related to public policy from their respective countries and its influence on ALS/MND clinical care and research. Experts provided information regarding governmental and private agencies related to support for patient care and research, assistive technologies, genetic testing, access to palliative and end-of-life care and Multidisciplinary clinics. Legal status of advance directives, palliative sedation, assisted death and euthanasia was also provided by ALS/MND experts for their respective countries.

Results:
Major disparities exist among countries which in turn reflect ALS/MND public policy and its varying ability to support patient care and research in addition to multiple other areas of importance. These areas include, but are not limited to, public perception and awareness of ALS/MND, caregiver support, quality of life for patients and caregivers, economic support for disabled patients and high quality end-of-life care.

Major disparities in multiple domains of public policy were noted among WHO-designated high-, middle- and low-income countries. Several countries had virtually no public awareness of ALS/MND and very limited means to support patient care and little if any for research. Numerous factors influencing public policy include economics, culture, religious beliefs, and societal values which are often inter-related.

Discussion:
This international survey from ALS/MND experts representing twenty-one countries provides valuable insights into factors which influence the development of ALS/MND public policy. These factors vary greatly among WHO-designated high-, middle- and low-income countries. Major disparities exist in providing patient care and funded ALS/MND research.

Information obtained from this international, cross-cultural survey serves as a platform to develop innovative and collaborative approaches to formulate optimal ALS/MND public policy, while taking into account inherent economic limitations for many countries. The additional influence of cultural and religious factors are significant and must be taken into account.

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Acknowledgements:
The authors thank the numerous experts who gave generously of their time while providing key insights into their respective country's ALS/MND public policy.

CMS-19: Nationwide implementation and evaluation of an e-health innovation for personalized care for patients with amyotrophic lateral sclerosis (ALS Home monitoring & Coaching) using participatory action research

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

Background:
ALS Home monitoring & Coaching is eHealth care for patients with ALS/MND based on home monitoring of functioning, body weight and well-being. Patients transfer these monitoring data through a (web)app to a dedicated ALS team member who, in consultation with the ALS team, takes action if necessary, e.g. giving advice, providing information, and planning consultations. ALS Home monitoring & Coaching is implemented as usual care in the University Medical Center Utrecht with high adoption and adherence rates, and positive user experiences for both patients and healthcare providers (Helleman et al., 2020).
Aim:
This ongoing study aims to implement this e-health innovation nationwide, starting with 10 multidisciplinary ALS teams spread across The Netherlands. This research focuses on the implementation process, barriers, facilitators and strategies for implementation and the feasibility, acceptability, and usability for patients and healthcare professionals.
Methods:
A Participatory Action Research approach is used for implementation, with researchers working together with rehabilitation specialists, allied health professionals, managers, patients and informal caregivers at each location. The implementation in each ALS care team consists of three stages with a total duration of approximately 5-6 months: 1) exploring/identifying expected barriers and facilitators; 2) developing strategies to address expected barriers and transforming these into actions; and 3) pilot phase in which ALS care teams include 10 patients to test and execute their implementation plans and provide care with ALS Home monitoring & Coaching. After three months the implementation outcomes and user experiences are evaluated using both qualitative (e.g. focus groups) and quantitative methods (online questionnaires).
Results:
Preliminary results show that the ALS teams are convinced of the importance and the benefits of ALS Home monitoring & Coaching. The reported barriers are related to innovation characteristics (lack of integration into the electronic health record), characteristics of the healthcare providers (new roles and responsibilities; beliefs that eHealth would disrupt the delivery of care; perceived threats to patient-professional relationship), and the organizational context (compatibility with workflows; concerns about reimbursement). Evaluation of the first pilot study showed a high adoption rate (100%) and positive user experiences. Patients and healthcare providers rated ALS Home monitoring & Coaching on average with an 8.0 (SD 1.2) and 7.4 (SD 1.3), respectively.
Discussion:
Preliminary results show that ALS Home monitoring & Coaching can be successfully implemented in different settings (ALS teams in rehabilitation centers, hospitals and university medical centers) by engagement of key stakeholders using a Participatory Action Research approach. In addition, the program seems to be feasible, acceptable, and usable for patients and their healthcare providers.

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CMS-20: Palliative care for ALS/MND - a European survey of practice

Professor David Oliver[1], Professor Gian Domenico Borasio[2], Professor Patrick Cras[3], Professor Marianne de Visser[4], Dr Nilay Hepgul[5], Professor Stefan Lorenzl[6], Dr Ludo Vanopdenbosch[7], Dr Simone Veronese[8], Professor Raymond Voltz[9]
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Live Poster Session B, December 10, 2020, 5:10 PM - 5:50 PM

Background:
Palliative care services have been closely involved in the care of people with ALS/MND from the 1960s, with patients admitted to St Christopher's Hospice soon after it opened in 1967. A survey in the UK in 2000 showed that over 75% of specialist palliative care services were involved in ALS/MND care, but often only in the later stages (1)

Introduction:
In 2019 a survey across Europe undertaken by the European Association for Palliative Care (EAPC) Reference Group on Neurology and the European Academy of Neurology Scientific Panel on Palliative Care showed that ALS/MND was one of the commonest diagnoses for close collaboration with 63% of palliative care specialists and 70% of neurologists reporting strong or moderate collaboration (2). This survey has looked in greater depth at the experiences of palliative care in Europe.

Methods:
An online survey will be undertaken of palliative care involvement and experiences of collaboration with neurology across Europe, starting in October 2020. Palliative care participants will be recruited through the EAPC Reference Group on Neurology, the EAPC website and blog, at the EAPC Research Congress and through palliative care organisations in Europe.

Results:
The results are being collected and will be available for this Symposium. Details of the extent of collaboration with neurology services, the methods of collaboration, the members of the multidisciplinary team, the barriers to collaboration, the expertise of the palliative care team, the areas that palliative care are most able to help, the involvement in non-invasive ventilation and tracheostomy ventilation and any changes seen during the COVID19 pandemic.

Discussion:
It is envisaged that there will be evidence of collaboration across Europe but this will vary both within countries and between countries. The timing of involvement is expected to vary and may be concentrated in the later stages of the disease progression, although earlier involvement has been suggested (3). It is hoped that the results will enable recommendations to be developed to foster further collaboration, and to enable the development of improved care for people with ALS/MND throughout the disease progression.

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CMS-21: Predictors of self-perceived health worsening over COVID-19 emergency in ALS

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

Background:
The international outbreak of Corona Virus Disease 2019 (COVID-19) has caused a health system emergency worldwide, calling for restrictive measures to prevent further spread. While COVID-19 represents a severe health threat for everyone, many neurological patients are at increased risk compared to the general population. Among them, patients with respiratory insufficiency from neuromuscular weakness, such as patients with ALS, are likely to be at higher risk of severe COVID-19 infection. Notably, besides a greater vulnerability in case of infection, ALS patients are also more vulnerable to the indirect effects of a quarantine since they are highly dependent on other figures (e.g. caregivers, neurologists, physical therapists, and others) to manage their chronic disease and maintain a good mental and physical health.

Objective:
We aimed at exploring self-perceived mental and physical health status over COVID-19 emergency in patients with ALS, and at investigating its main predictors among demographic and disease-specific features, as well as clinical care changes.

Methods:
A web-based questionnaire investigating self-perceived anxiety, depression, and clinical worsening, as well as clinical care changes over COVID-19 emergency was administered to ALS patients currently followed at San Martino Hospital. Ordinal and logistic regression analyses were applied to identify significant predictors of self-perceived health status.

Results:
57 ALS patients completed the questionnaire. 35.08% of ALS patients reported anxiety symptoms, 36.84% depressive symptoms, and 35.08% a clinical worsening. Concerning clinical care changes, only 3.50% of ALS patients experienced problems in retrieving medicines, while 63.15% of patients had to cancel exams and/or visits. 75.44% of respondents were following a rehabilitation therapy at the time of the outbreak and, among them, 79.06% had to suspend it over the quarantine period. Significant predictors of anxiety symptoms severity were female gender (P=0.04), greater motor impairment (lower ALSFRS-r score, P=0.02), more aggressive disease course (higher ALSFRS-r decline per month, P< 0.001), and rehabilitation therapy suspension (P=0.02). The only predictor of depressive symptoms severity was a more aggressive disease course (higher ALSFRS-r decline per month, P=0.05). Significant predictors of clinical worsening were shorter disease duration (P=0.05) and exams and/or visits cancellation (P=0.005).

Discussion:
Clinical care changes over COVID-19 related quarantine exerted a significant impact on self-perceived health status in patients with ALS, particularly in those in the earliest disease phases and with a more aggressive disease course. These findings have potential to improve disease-oriented, personalized patients management in the next phase.

Acknowledgements:
We would like to thank patients for participating in the study. No specific funding was received for this work.

CMS-22: Recruiting MND patient research participants: I thought we would have more.

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

Introduction:
LOROS Hospice provides a service to all people with MND in Leicester, Leicestershire and Rutland from the point of their diagnosis and research is an integrated aspect of the services. We wished to offer patients the opportunity to participate in the COMMEND study, a randomised controlled study of commitment and acceptance therapy. Despite a first stage feasibility study no guidance was available to inform the likely number of eligible participants.

The aim of this work was to identify the potential number of patients that could be approached about the study and understand the differences between the total population and this sample and its implication for study feasibility assessments.

Methods:
The notes of all patients with MND who were registered at the Hospice were identified and mapped to the inclusion and exclusion criteria of the COMMEND study.

Results:
70 patients were identified of whom only 19 (27%) were eligible for the COMMEND study. The largest number (33%) were excluded because of receiving nutritional and/or ventilator support. A further 20% were excluded because of cognitive issues and 6% did not speak English as a first language.

Conclusions:
That less than a third of patients were eligible for this relatively straightforward interventional study was a surprise to us. It is a well-known aphorism that potential participants disappear as soon as a study opens. This work provides a screening analysis which informs realistic feasibility assessment and more nuanced understanding of the MND patient population. As LOROS provides care to all patients in the geographical area these findings should be generalizable.

CMS-23: Refining items for a preference-based, ALS specific, health related quality of life scale

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

Background:
Disease specific preference-based health-related quality of life (HRQL) measures provide a description of the impact of ALS on HRQL, assist with resource planning, and incorporate patient's perspective of care[1,2].
Existing preference based HRQL measures used in ALS are generic, and may not include domains that are important to people with ALS (PALS)[1]. There is a need to develop an ALS specific preference based HRQL measure that is co-produced with PALS.

Objective:
This study aimed to develop the initial items of a PALS preference-based ALS HRQL scale (PB-ALS Scale) based on previously generated domains.

Methods:
Demographic details including age, ALSFRS-R score, etc. were collected from each participant. PB-ALS Scale items were refined using survey methodology to assess importance ratings and item independence. Median importance was calculated for each item. A median value of 3 (very important) was required for the item to remain. Items that were highly correlated with other items (Spearman's correlation coefficient >0.7) were removed.

Cognitive debriefing interviews were used to refine the survey-developed PB-ALS Scale items. Zoom, telephone, or email interviews were conducted to accommodate PALS' preferences and communication needs.

Participants provided feedback (wording clarity, response options, recall period, etc.) on 3-5 randomly selected PB-ALS Scale items. Items were considered finalized when three sequential participants approved the item without any revisions[3].

Results:
Thirty-four participants (n=16 female; ALSFRS-R range = 1-48) from Eastern, Central and Western Canada completed the survey. Eighteen participated in the cognitive interviews (n=8 female, n=10 male). Four items were highly correlated with one or more other items or were not rated important by participants and were removed. The final PB-ALS scale includes eight items with four response options addressing the following domains: recreation and leisure, mobility, interpersonal interactions and relationships, eating and swallowing, handling objects, communicating, routine activities, and mood.

Next Steps:
In phase three, the eight finalized items of the PB-ALS Scale items will be translated into French and patient preferences will be used to develop a scoring algorithm.

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3. Kuspinar A, Bouchard V, Moriello C, Mayo NE. Development of a bilingual MS-specific health classification system: The preference-based multiple sclerosis index. Int J MS Care. 2016;18(2):63-70.

Acknowledgments:
Thank you to the PALS who participated and the research team members for their support. This study was funded by a 2018 ALS Society of Canada Project Grant and the James and Jeanie Brown ALS Research Fund from the University Hospital Foundation, Edmonton, Alberta.

CMS-24: The Association of Bulbar Impairment and Communicative Participation in ALS

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

Background:
The importance of social connection to mental health, physical health, and quality of life is well-documented. While complex and multi-determined, the foundation of strong social connection is successful communication interactions. Therefore, individuals with communication disabilities are at risk for social disconnection. Bulbar involvement in ALS often results in dysarthria, a neuromotor disorder of speech execution reflecting disruption to the speech subsystems (respiration, phonation, resonation, articulation). The onset of speech impairment frequently leads to reduced speech naturalness, clarity, and restricted communicative participation (the use of communication in daily life situations). Though a few studies have demonstrated a relationship between person-report measures of speech function and communicative participation by PALS, the contribution of specific speech features is unclear. This gap hinders effective approaches to monitoring functional speech outcomes and to promoting social connection.

Objective:
The study explores the association of measures of quantitative motor speech decline in ALS (speaking rate, pausing, articulation) to communicative participation. Our aim is to understand the clinical importance of quantitative motor speech analysis and to identify speech therapy treatment targets.

Methods:
Data were collected from 26 PALS using the Beiwe smartphone research platform. On the Beiwe app, participants completed 1) surveys - the Communicative Participation Item Bank Short Form (CPIB) and the Revised ALS Functional Rating Scale (ALSFRS-R), and 2) audio-recordings - a reading passage and speech diadochokinetic task (repetition of /puh-tuh-kuh/). Speech acoustic measures (speaking rate, articulation rate, percent pause, articulatory precision, voice-onset time) were extracted offline. Communicative participation was compared between PALS with and without bulbar signs (based on ALSFRS-R bulbar subscale score). Bivariate correlations between the CPIB and speech/bulbar scores were conducted. Multiple linear regression models will be developed to examine the unique contribution of each of the variables to the prediction of CPIB scores.

Results:
CPIB scores were significantly lower for PALS with self-reported bulbar signs ($p<.001$). Significant correlations ($p<.05$) were observed between the CPIB and a number of acoustic speech features (speaking rate, articulation rate, articulatory precision, voice onset time). However, percent pause time was not significantly correlated with the CPIB score ($p=0.11$). ALSFRS-R speech (Q1) and bulbar subscale scores were significantly correlated with the CPIB ($p<.001$), while the respiratory subscale was not ($p=0.22$).

Discussion:
Our preliminary findings indicate that communicative participation is significantly restricted in PALS with bulbar signs, and that speech features related to articulator movement, but not respiratory function, are related to communicative participation. Further investigation is warranted to clarify the clinical impact of motor speech decline, and to facilitate the management of communicative participation and social connection.

Acknowledgments:
This study was supported by the ALS Association, the Winthrop Family Fund for ALS Science at Massachusetts General Hospital, NIH grant DP2-MH103909, and NIH-NIDCD K24DC016312.

CMS-25: The impact of bulbar and cognitive deficits on coping with ALS and navigating healthcare services

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

Background:
Cognitive deficits in the presence of bulbar symptoms may be exaggerated in patients with ALS(1). These impairments can hinder timely delivery of speech-language pathology (SLP) management practices, which typically use patient reports with clinician judgements(2). Despite clinical efforts to provide support, patients and caregivers continue to struggle with managing bulbar and cognitive deficits(3). Multiple calls to action have been made to ensure clinical management is informed by patient and caregiver's priorities(4).

Objectives:
To describe the experiences and challenges of ALS patients with bulbar and cognitive deficits to inform best practice guidelines for improving SLP services.

Methods:
A thematic narrative analysis was applied to data transcribed from a qualitative interview. 6 ALS patients (2 Females, 4 Males, mean age = 55 years) and their caregivers participated in semi-structured interviews discussing their experiences on coping with ALS and SLP services. Patients were recruited based on documented bulbar disease and cognitive deficit, defined as below cut-off scores on a neuropsychological battery. All patients had significantly reduced speech intelligibility. Four patients presented with mild cognitive impairment (MCI); two patients met the criteria for frontotemporal dementia (FTD).

Results:
Four themes were identified emphasizing the need for education, advocacy, personalized care management, and caregiver support. Caregivers who had partners with FTD expressed substantial difficulties supporting their partners and advocated for timely and more personalized SLP interventions. Patients with MCI got by using cognitive strategies independently, whereas caregivers of FTD patients implemented cognitive strategies taught by SLPs. Patients with FTD and their caregivers were found to have more difficulties with emotionally coping with the changes in ALS.

Discussion:
Patient perspectives serve to raise awareness to those with lived experiences and remind SLPs of their role to better support individuals living with ALS across the cognitive continuum. SLPs are encouraged to (i) provide more education on bulbar disease prognosis and symptom management, (ii) support patient advocacy, (iii) focus on providing more personalized interventions, and (iv) develop means of caregiver support throughout ALS progression.

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Acknowledgements: We would like to thank all participants for taking part in the study. Funding for this study was provided by National Institutes of Health from the NIH-NIDCD R01DC017291 grant, the ALS Society of Canada and Brain Canada from the Hudson Translational Team Grant, and the TD Bank Financial Group from the KITE—Toronto Rehab's TD Graduate Scholarship for Students with Disabilities.

CMS-26: Triage of Amyotrophic Lateral Sclerosis patients during the COVID-19 pandemic: an application of the D50 model

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

Background:
Amyotrophic Lateral Sclerosis (ALS) is a progressive neuromuscular disease the management of which requires the continuous provision of multidisciplinary therapies and prescient patient counselling. Owing to the acute COVID-19 pandemic, effectively reaching Germany in March 2020, regular contact with ALS patients at our center was severely restricted and patient care was at risk of the indefinite delay of supportive therapies.

Objectives:
We were therefore dependent on a triage system for the patients with ALS, in order to avoid in- and out-patient consultations as far as possible whilst preserving the principles of advanced care planning as well as possible. We used the D50 disease progression model for this purpose and were thus able to identify a prospective cohort with high disease aggressiveness (D50 < 30 months) [1-3].

Methods:
We identified a group of 75 high aggressive patients, 102 had lower disease aggressiveness. We offered active follow-up visits, either via telephone or on-site, depending on their disease-specific needs and abilities. We describe here the procedures, obstacles and results of these prescient efforts during the restrictions caused by COVID-19 in the period between March and June 2020.

Results:
Twenty of the 75 highly aggressive patients (26.7%) had already died in the 1 year preceding March 2020, whilst in the group of lower aggressive patients only 5 people had died (4.9%; $p < .001$). Thirty-seven of the highly aggressive patients responded to our active offer for follow-up. During the regarded 3 months (March-June 2020), 2 highly aggressive patients received a gastrostomy, 4 were initiated with non-invasive ventilation (NIV) and another 8 patients were prepared for the timely initiation of NIV. We could show that a comparable amount of advanced care was induced in a retrospective cohort within a similar time period 1 year prior to the COVID-19 outbreak. The total ALSFRS-R for the patients in the high aggressive group significantly declined from a median of 34 to 28 points ($p < .001$).

Discussion and conclusions:
Our workflow to identify high-risk patients via the D50 model can be easily implemented and integrated within existing centers. It helped to maintain high quality of advanced care planning for the patients with ALS treated at our center.

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1 Steinbach R, Prell T, Gaur N et al J. Clin. Med. 2020; 9(9), 2873.
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This work was supported by the German Bundesministerium für Bildung und Forschung (BMBF) and Deutsche Forschungsgemeinschaft (DFG). Support was also received from the Motor Neurone Disease Association (MNDA) and Deutsche Gesellschaft für Muskelkranke (DGM).

CMS-27: Using a Design-Build Approach to Improve International ALS Research News Access in Italy

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

Keywords:
design-build, research news access, clinical trials

Background:
Accessibility to accurate real time international ALS scientific research news in non-English languages is often a challenging endeavour for patients, families, and caregivers.

Objectives:
This presentation examines how a patient led, multi-disciplinary Italian ALS association works to improve the landscape of online international ALS research news availability in Italy.

Methods:
This project incorporates an innovative interdisciplinary design-build approach to improve information accessibility in the Italian ALS community by integrating patient feedback from the community into information resource development.

Results:
A partial summary of available features include: a user friendly multi-lingual website, Italy's first live international ALS clinical trial database, caregiver training course series, a general guide to Edaravone in Italy, ALSUntangled in Italian, news sharing on social media, and newsletters.

Discussion and Conclusion:
Preliminary results of these implemented initiatives have demonstrated increased community member engagement locally and abroad. Further work is necessary to improve existing information platforms in synchrony with increased domestic and international information exchange collaborations.

Acknowledgements:
We express our most sincere gratitude to the dedicated patients, families, caregivers, and volunteers from the Italian community for their participation.
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CMS-28: 70% of Australians want to die at home, but only 15% do. How the Medical Aids Subsidy Scheme Palliative Care Equipment Program (MASS PCEP) is meeting the needs of people with MND in Queensland, Australia.

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

Background:
In 2019 the Queensland Department of Health introduced Care in the Right Setting (CaRS) program. The CaRS program is dedicated to funding models of care, that support the provision of healthcare in the right setting and promote the appropriate and effective use of hospital services. One of the areas highlighted to receive pilot funding was Palliative Care. Medical Aids Subsidy Scheme (MASS) was successful in their submission to administer and coordinate a Palliative Care Equipment Program (PCEP). PCEP was launched in April 2020 and is funded until 30 June 2021. Palliative Care Australia has identified that only 15% of Australians die at home. Compared with other OECD countries such as New Zealand, Ireland and France, the rate of deaths at home in Australia is very low. The evidence shows that home-based palliative care is more cost effective, whilst improving quality of life. When palliative care is provided at home, these people are 87.5% more likely to remain in the community until death. MASS PCEP aims to assist to alleviate the financial and logistical challenges experienced by the family unit, through providing timely access to assistive technology. This is achieved through a coordinated and centralised hub with a single point of contact, with equitable delivery of services including rural and remote regions. There is no cost to a person to receive support from PCEP. To be clinically eligible, they must have a Palliative Care Medical Specialist confirm they are in their last six months of life and have a permanent residential address in Queensland.

Objectives:
The purpose of PCEP is to support a person to remain at home by providing:
continence aids and a home oxygen concentrator to help maintain quality of life;
daily living and mobility aids to assist in safely completing daily living tasks independently or with assistance.

Discussion/Results:
Qualitative outcomes will be presented in the form of a patient's story. George has truncal onset MND and had been unable to access the deck of his home for over 12 months. The impact PCEP has had on him and his family will be showcased. Quantitative outcomes will be presented such as: the number of applicants; the number of pieces and types of assistive technology; the duration of participation in PCEP. Also presented will be how PCEP has adapted and evolved to meet the needs of people living in the regional and remote areas of Queensland. People can be living over 2500 kilometres from the MASS office, based in the capital city of Queensland.

Reference:
Swerisseen, H. & Duckett, S.J. (2015) What can we do to help Australians die the way they want to? Medical Journal of Australia (1): 10-11. doi: 10.5694/mja14.01580

CMS-29: A survey on patients' disease perception and the impact of the COVID-19 pandemic on persons living with ALS in Malaysia

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

Background:
Knowledge on the understanding and management of Amyotrophic Lateral Sclerosis (ALS) in Southeast Asia is lacking. While there are a number of studies looking at patient experiences in high-income and Western countries, there is limited information about the experiences of ALS patients in low and middle income countries (LMIC). Specifically, the social and management aspects of persons living with ALS, including their use of complementary and alternative medicine (CAMs) and impact on finances are not well-understood. The current and ongoing COVID-19 pandemic would have also likely to have given rise to additional challenges and understanding its impact can inform medical providers and support groups on measures that can be implemented to better cater to patients' and caregivers' needs during crises.

Objectives:
The purpose of this study was to investigate patients' perception of their disease, its management and the impact of the COVID-19 pandemic on persons living with ALS in Malaysia.

Methods:
An online survey comprising of 40 questions was conducted which explored management of care and financial impact of ALS, as well as patient experiences due to the imposed restriction movement order during the peak of the COVID-19 pandemic in Malaysia.

Results:
Responses were received from 37/60 (62%) participants with ALS directly or through their caregivers. We found that the majority (89%) of Malaysian ALS patients were adequately informed about the disease, mainly through the internet. Interestingly, most patients (83.8%) relied on at least one form of CAMs and the majority perceived the treatment as having the potential to slow or stop disease progression. The disease added to their financial burden by reducing both patients' earnings (62.2%) and their caregivers' earnings (35.1%) and savings (54.1%). During the COVID-19 pandemic, two-thirds of patients (67.6%) were negatively impacted by the sudden disruption to their hospital appointments, rehabilitation sessions as well as reduced social interactions.

Discussion and Conclusion:
This study provided insight into patients' perception of their care and management of ALS in Malaysia which will facilitate in implementing changes that can improve care to persons living with this devastating illness.

Acknowledgment:
We extend our deep gratitude to our patients and their caregivers for participating in the survey. The study is funded by the Malaysian Ministry of Higher Education (FP043-2018A), ALS Association and Sydney South East Asian Centre. The authors declare no conflicts of interest.

CMS-30: Can Generic Preference-Based Measures Be Used to Capture the Health-Related Quality of Life of Individuals with Amyotrophic Lateral Sclerosis?: A Content Validation Study

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

Background:
Generic preference-based measures (GPBMs) are commonly used with people with Amyotrophic Lateral Sclerosis (PALS) to capture health-related quality of life (HRQL) and provide a single index score that can be used evaluate cost and clinical effectiveness of interventions1 in resource-constrained health systems. A measure's psychometric properties must be determined before use in a population to ensure values obtained are valid for interpretation and utilization2. The psychometric properties of GPBMs has not yet been established in ALS.

Objectives:
The objectives of this study were to: 1) examine convergent validity of the most widely used GPBM, the EuroQol 5 Dimension 5 Level (EQ-5D-5L), against the Patient Generated Index (PGI) in ALS; 2) assess content validity of seven GPBMs.

Methods:
Participants were recruited from three Canadian multidisciplinary clinical sites. The PGI and EQ-5D-5L were administered via online or hardcopy survey and scores compared for convergent validity. Domains nominated by participants as important to their HRQL, generated using the PGI, were classified using the International Classification of Functioning, Disability and Health (ICF), and mapped onto the seven most widely used GPBMs to determine percent content coverage.

Results:
Fifty-two PALS (N=28 female; mean age = 61.3 ± 11.6 years; mean time since diagnosis = 3.5 ± 2.9 years) participated. A moderate correlation (Pearson's r=0.52) between the PGI and EQ-5D-5L (53% content coverage) was found. The top three ICF domains identified in the PGI were: recreation and leisure, lower limb mobility, and interpersonal relationships. No one widely used GPBM covered all areas of HRQL identified as important. The Quality of Well-Being Self-Administered scale had the highest content coverage (87%). The Health Utilities Index 3 had the lowest (33%). Only two domains identified as important in the PGI were included in all GPBMs.

Discussion:
The majority of GPBMs included about half of the domains PALS identified as important. The most commonly used GPBM in ALS, the EQ-5D-5L, underestimated the impact of ALS on HRQL. Some domains identified using the PGI were not covered by GPBMs, suggesting the need for an ALS specific preference-based measure to better reflect the health concerns of this population and support appropriate resource allocation to support the needs of PALS.

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CMS-31: Management of Vulnerable Patients During the COVID 19 Pandemic

Dr Deirdre Murray[1], Mr Colm Peelo[1], Ms Mairead Campbell[2], Ms Eithne Cawley[3], Dr. Amina Coffey[1], Ms Sarah Coleman[2], Ms Bernadette Corr[2], Ms Lesley Doyle[2], Dr. Eoin Finnegan[1], Ms Olivia Grogan[2], Ms Louise Hennessey[3], Mr Mark Heverin[1], Ms Katie Kinsella[3], Ms Teresa Leahy[5], Dr. Sinead Maguire[2], Ms Kitty McElligott[2], Ms Aisling O' Reilly[2], Ms Fidelma Ruttledge[3], Prof Aisling Ryan[4], Dr. Marie Ryan[1], Ms Roisin Vance[2], Prof Orla Hardiman[2]

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

Background:
The rapidly progressive nature of ALS requires timely multidisciplinary (MDT) intervention. In addition to physical disability, cognitive and behaviour changes cause significant burden for carers. The Covid-19 pandemic severely disrupted the established model of care, based on a combination of regular attendance at specialised MDT clinics and home visits, which include set-up of non-invasive ventilation (NIV). During the first wave of the COVID-19 pandemic in Ireland (March-July 2020), the National ALS service combined in-person and telemedicine services and enhanced home visiting. National planning meetings utilised video conferencing. The aim of this audit is to evaluate the impact of Covid-19 on the service.

Method:
The provision of services prior to Covid-19 was compared with that during the 'first wave' (March to July 2020) using key statistics, including mortality, collected by the National MND Register.

Results:
Between 20th March¬ and 1st July 2020 there were 213 clinic visits compared with 198 during the same period in 2019. A similar number of new patients (38 vs 32) were seen, with 97% being face-to-face visits. 72% of return patients (n=126/175) elected for remote consultation. All patients were triaged in advance by phone call. Respiratory monitoring utilised home-spirometry with video guidance or in-clinic SNIP, with 64 measurements (50 remote FVC), compared with 109 in 2019. A similar number of home visits were conducted (18 per week), with enhanced precautions, including NIV set up (n=21). A new physician home visit service commenced. Although patients seen within-clinic by the MDT reduced from 107 in March to July 2019 to 46 in 2020, the MDT adapted, with 49 consultations by the Dietitian, 34 by the Occupational Therapist (44% remotely), 93 by Physiotherapy (66% remotely).

During this period of the pandemic, 2 (0.05%) of ALS patients developed COVID-19, and one died from associated complications. Presentations to the Emergency Department were reduced sixfold. Mortality rates from ALS did not change during this period.

Discussion:
The Covid-19 pandemic resulted in a rapid transformation in the MDT MND services. Essential services were maintained to a significant degree, using video and telephone consultations and enhanced home visits. The demands in the acute hospital setting including redeployment and limited access to community services were significant challenges. Delivering services in a safe manner required significant planning. In preparation for a second wave, additional remote monitoring is being implemented using a modified version of the Sheffield Telemedicine in MND (TiM) application(1). Evaluation of this service change will be conducted through patient consultation to ensure positive changes are maintained and gaps identified.

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CMS-32: Neck weakness in Motor Neurone Disease: An Insight into Head Support

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

Background:
Neck weakness is a symptom of Motor Neurone Disease (MND), resulting in the head dropping forward or to the side, due to gravity and the weight of the head (Gourie-Devi et al, 2003). If it is not managed appropriately, this can result in issues with breathing, swallowing, communication, skin care, visual awareness as well as abnormal posture, which can lead to contractures and pain (Alghadir et al, 2017; Oliver, 2019).
There is currently a paucity of information available regarding neck weakness in MND and as a result there are no recommendations in the NICE guidelines to assist with the management of this problem.

Objectives:
To explore the characteristics of neck weakness in MND and the different types of head support strategies used in MND.

Method:
Cross sectional survey, using a consecutive cohort from a multidisciplinary centre at the Statewide Progressive Neurological Service (Calvary Health Care Bethlehem, Melbourne, Australia). Participants with a diagnosis of MND with neck weakness answered an online questionnaire (17 items) about their neck weakness and the various strategies that they currently used for head support.

Results:
The questionnaire was sent out to 40 patients and 33 responded. The median time for the onset of neck weakness from time of diagnosis of MND was 7 months (IQR = 3-23). All participants reported that neck weakness affected their quality of life to some extent. The majority of participants reported current use of 2 or 3 different types of head support strategies. The most commonly used strategy was using tilt/recline function (87.9%) followed by soft neck collar (63.6%) and aspen vista collar (21.1%). Half of those who use a neck collar, reported an inability to wear it for as long as they need citing several reasons for this.

Discussion:
This study demonstrated the variability in neck weakness characteristics and head support strategies that people with MND use. It highlights the importance of an individualised approach to management of neck weakness and head drop in MND. Exploration of how neck weakness and head support strategies changes over the course of the disease would be an important study to pursue to further research in this area.

Acknowledgments:
We would like to thank our patients and the Physiotherapy department at CHCB.

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Oliver DJ. Palliative care in motor neurone disease: where are we now? Palliat Care. 2019; 12: 1178224218813914.

CMS-33: People living with ALS and their caregivers' input into drug development in Europe

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

Background:
There is an emerging focus on patient and caregiver engagement in determination of clinically meaningful outcomes, and input to key areas of drug development. The European Medicines Agency (EMA) is considering methods to better incorporate patient and caregiver input into regulatory review processes.
The IMPACT European survey of people with amyotrophic lateral sclerosis (ALS) and caregivers is collecting data on the overall burden of disease, psychological distress and the loss of function over the course of the disease.

Objective:
To survey European ALS patients and caregivers across 9 countries. The survey captures details on patient and caregiver perspectives across the illness journey. Patients and their caregivers may have different concerns regarding the burden of disease, and comparisons of perspectives will be analyzed. Results of the European survey will be viewed in comparison with results from a similar 2017 US survey.

Methods:
A steering committee was established, consisting of industry partners, clinical and methodological experts, with input from patients and caregivers. The survey materials used in the US were adapted for use in Europe. Most questions were directly comparable, with relevant amendments for local contexts. Recruitment of patients and caregivers was carried out with the partnership of European Network for the Cure of ALS (ENCALS) and advocacy groups in each country. The survey questionnaires – one for people with ALS, one for current caregivers and another for bereaved caregivers – were hosted on a Qualtrics platform. Ethical approval was received from Trinity College Dublin, participant confidentiality was agreed, and all relevant GDPR regulations were followed during survey processing, analysis and dissemination of findings.

Results:
Descriptive statistical analysis of European patient and caregiver data, and free text analysis of open-ended responses. The survey results will also be analyzed in conjunction with the results of a similar survey carried out in the United States. Findings from the US survey included a more detailed understanding of the functional burden of living with ALS and the identification of a number of clinically meaningful symptoms beyond muscle weakness that could be the focus for future therapies

Conclusions:
The IMPACT European patient and caregiver surveys will provide information on the disease burden from the perspectives of people living with ALS and those providing care. and provide guidance into drug development processes.

Acknowledgements:
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CMS-34: Physical Activity in People with Motor Neuron Disease: PASE as a Measuring Tool

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Background:
The reduced activity that occurs as a result of chronic disease and disability can lead to a cycle of secondary deconditioning, physical deterioration and further subsequent reductions in activity. While the consequences of inactivity and exercise is well documented in neurological conditions such as stroke, multiple sclerosis and Parkinson's disease, less is known about the exercise behaviour of people with MND. To date, there have been no studies looking at how the disease affects physical activity and exercise participation by people with MND.

The PASE (Physical Activity Scale for the Elderly) is a questionnaire designed to measure physical activity participation in older adults. It has been used in numerous studies exploring physical activity in people with progressive neurological conditions such as Parkinson's disease (PD).

Objective:
This study aims to investigate the usefulness of the PASE as a tool to measure physical activity in people with MND. It will explore the types of activities people with MND are choosing to continue participating in, and the factors that influence physical activity participation as the disease progresses.

Methods:
A prospective, observational study (Registered: ACTRN12618000889257) was undertaken at a multidisciplinary MND clinic in Victoria, Australia. All patients with MND who were still ambulant (with or without assistance) were recruited to participate in the study.

Demographic data, PASE, FVC, ALSFRS-r and falls history were taken for analysis. Information about the types of activities people with MND choose to do were extracted from the PASE to observe for common themes and categorised. If participants acknowledged that there were barriers to physical activity participation, fixed choice options were presented to best explain the barrier and response frequency was calculated.

Results:
There were significant large positive correlations between PASE and ALSFRS-R total scores (rho = 0.607; $p<0.001$). Both univariate and multivariate regression analyses showed that disease severity (ALSFRS-R total score) was the strongest predictor of PASE score.

71% of participants participated in walking activities around their home and 51% performed housework. Only 10% participated in group exercises and 12% participated in sporting activities. Fatigue was the biggest barrier to exercise participation in this population.

Conclusion:
The PASE questionnaire is a quick and easy tool that can be used to measure self-reported physical activity in ambulant adults with MND. It correlates well with the current gold standard of functional outcome measure in this population, the ALSFRS-r. Physical activity participation in this population is most strongly influenced by disease severity, particularly walking ability and upper limb function.

When it comes to physical activity choices, walking outside the home was the most common answer followed by house work. Participation in exercise groups and sports were the least frequent response.

CMS-35: Safety information of patients from ongoing post-marketing surveillance, evaluating the real-world efficacy and safety of edaravone for amyotrophic lateral sclerosis patients in Japan (SUNRISE Japan)

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Background:
Edaravone, a free-radical scavenger developed as a neuroprotectant agent, was approved for the treatment of patients with amyotrophic lateral sclerosis (ALS) in various countries from 2015 to 2020 (Japan, June 2015; South Korea, December 2015; United States, May 2017; Canada, October 2018; Switzerland, January 2019; China, July 2019; and Indonesia, July 2020). The approvals were based on efficacy and safety data from patients with a definite or probable ALS diagnosis[1]. Edaravone demonstrated statistically significant efficacy in slowing the progression of ALS, as assessed by scores on the ALS Functional Rating Scale-Revised (ALSFRS-R). However, to date, other end points, such as survival time or time to tracheal intubation, have not been assessed. Therefore, this ongoing study, SUNRISE Japan, aims to obtain real-world data about the long-term efficacy, including survival endpoints, and safety of edaravone in ALS patients in Japan.

Objectives:
To assess the safety profile of edaravone through post-marketing surveillance for patients with ALS.

Methods:
Overall, more than 800 ALS patients who were edaravone treatment−naïve have been enrolled in the surveillance program and will be followed up for 5 years. Safety assessments include adverse events up to 1 year. Efficacy assessments include duration of survival and duration until invasive tracheal intubation is needed up to 5 years; clinical events such as introduction of tube feeding, gastrostomy, and intermittent noninvasive ventilator assistance up to 1.5 years; and ALSFRS-R scores up to 1.5 years. The survey is being conducted in accordance with Japan's Ministerial Ordinance on Good Post-marketing Study Practice.

Results:
Of the data summarized as of April 3, 2020, safety information on 800 patients included in the safety analysis is reported. Adverse drug reactions occurred in 97 patients (12.13%), and serious adverse drug reactions occurred in 30 patients (3.75%). The most common serious adverse drug reaction was "Hepatic function abnormal" which occurred in 6 subjects (0.75%). In this report, no unexpected safety signals were seen, nor any inconsistencies with the clinical trials.

Discussion:
This program may be useful in understanding longer-term effects of edaravone in the treatment of ALS in Japan and across the globe, as well as serving as a model for other long-term surveillance studies in neurodegenerative diseases.

References:
1) Writing Group on behalf of the Edaravone (MCI-186) ALS 19 Study Group. Lancet Neurol. 2017;16:505-512.

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We would like to thank the patients and investigators for taking part in this post-marketing surveillance. The study was funded and conducted by Mitsubishi Tanabe Pharma Corporation, Tokyo, Japan (MTPC). Kenta Y, KI, Kengo Y, MM, YK and SY are employees of MTPC.

CMS-36: Telehealth in Motor Neuron Disease (TiM): Rapidly developing and implementing a digital, remote monitoring service during the COVID-19 pandemic.

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Background:
Attendance at specialist MND clinics has been reported to improve survival for people with MND (PwMND). However, traveling to these centres can become difficult or impossible as the condition progresses. Additionally, appointments often occur at regular fixed intervals and thus are not responsive to the needs of PwMND or their carers. Therefore, some PwMND decline rapidly and go several weeks without being seen by their specialist clinical team despite a desperate need. Whereas other PwMND may attend their regular appointment after not having declined since their last clinic, and waste valuable days where they could be with their family instead.
To overcome these problems, Sheffield Institute for Translational Neuroscience developed a telehealth in MND (TiM) system which invites PwMND and their carers to answer weekly questions regarding how they are coping which can be reviewed by the MND care team. Healthcare professionals (HCPs) can then respond through the system, arranging a follow-up meeting, sending an informational leaflet, or reassuring message. Since the COVID-19 pandemic, the ability to provide remote care has become even more important. Therefore, to help continue to support those under our care, the team rapidly increased the development and implementation of TiM. This poster will describe the progress of the project in addition to some of the key lessons that have been learnt as a result of developing a novel service during an international pandemic.

The TiM system went live and begun inviting PwMND and their carers in May 2020. Currently, 30 people are using the system regularly and several developments have been implemented as a result of user feedback. Although several patient and public involvement (PPI) groups were arranged, due to COVID-19, these were unable to proceed, and the team have had to adapt to developing the system and making iterative changes based upon more informal feedback. Regular meetings with the HCPs monitoring the system have been essential, with a keen understanding of how TiM can supplement the existing services which are provided. Additionally, we have offered PwMND the chance to provide feedback through the TiM system, providing greater information to those invited to make it clearer as to the aims of the service.
Lastly, we have designed and scheduled two research projects which will fully capture the experiences of those engaging with TiM to help facilitate further development and provide useful lessons for other, similar services.

Discussion:
Through iterative, informal PPI feedback and system development, we have implemented a novel system to provide support for PwMND and their carers during COVID-19:

Acknowledgements
We would like to acknowledge MND Scotland for funding the project, the Sheffield MND Care and Research Centre, and everyone who has provided their time to help us develop the system.

CMS-37: Telemedicine experience of an Italian ALS tertiary Center during COVID-19 pandemic

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Objective:
To describe the telemedicine experience of an Italian ALS tertiary Center during COVID-19 pandemic.

Methods:
All visits scheduled between 6th March and 6th April 2020 were considered for telemedicine. Additionally, the Center maintained the possibility for patients to phone call specialists. A few days after each contact, patients and caregivers were surveyed for assessing their satisfaction with the service received.

Results:
A total of 144 controls were scheduled during the study period and consisted mostly of neurological or psychological visits (139, 96.5%). One hundred thirty-nine (96.5%) visits was performed as telemedicine and mostly via phone call (112, 80.6%). Three (2.1%) visits were considered as urgent and maintained as outpatient care. Patients were globally stable with respect to the previous visit (median Δ-ALSFRSr total score 1, IQR 0-4). Many requests of contact were addressed at getting information about the scheduled visits or examinations (45, 43.3%). Globally, patients and caregivers were satisfied of the telemedicine service. However, the majority (85, 65.9%) would globally prefer a face-to-face visit.

Conclusions:
We were able to rapidly adapt our assistance to the social restrictions due to COVID-19 pandemic. Such organization was fully appreciated by our patients and caregivers and could be a solution for the future, also regardless of the COVID-19 spreading.

CMS-38: The need to consider 'temporality' in person-centred care of people with motor neurone disease

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Aims and objectives:
The overall aim of this presentation is to provide practical insight into the way that professionals caring for a person with motor neurone disease (MND) can recognise, respect and respond to that person's temporality; i.e. the person that they have been, that they are now, and that they will be in the future.

Background:
MND is an umbrella term for a group of four rare, devastating neurodegenerative terminal diseases of middle/later life. Previously we have acknowledged the importance of different time-periods in the trajectory of MND as an illness, for example, during the diagnosis stage through to end of life and decision making at that time. Living with MND can cause anxiety at all stages of the disease trajectory especially as it can be difficult for people living with MND to communicate their desires and concerns to professionals and carers. It is important that professionals continue to provide holistic care throughout the illness trajectory and the aim of this presentation is to explore past research about caring for someone with MND in relation to the concept of person-centred care.

Method:
The presentation is based on the concatenated exploration of the findings of a hermeneutic phenomenological project. Thus, this discursive presentation links elements/studies which have been published previously to develop a model of person-centred care for people with MND which recognises and respects their temporality.

Conclusions:
We suggest MND has a significant impact on a person's lifeworld. The proposed person-centred care model focuses on understanding (interpreting) a person in a wider temporal frame, and beyond the context of their illness. The expected collaborative outcomes are that: a person is acknowledged as more than a 'patient with MND' and that a professional is providing person-centred care based on individuality of the person, through a temporal lens. This requires a collaborative approach between the person, others, and professionals. Such person-centred care, focused on individuality, may prevent a person experiencing life in crisis and suffering towards end-of-life.
Relevance to clinical practice: Our project findings provide insight into the need to consider temporality when caring for people living with MND. This is important as it can support a change in person-centred care and culture for people diagnosed and living with MND.

CMS-39: Treatment Persistence Among Amyotrophic Lateral Sclerosis Patients Receiving Intravenous Edaravone: Results From a US National Infusion Center

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Background:
Intravenous (IV) edaravone (Radicava®; Mitsubishi Tanabe Pharma Corporation, Tokyo, Japan) was approved by the United States (US) Food and Drug Administration (FDA) for the treatment of amyotrophic lateral sclerosis (ALS) in May 2017 and became available to US health care providers in August 2017. The approval was based on a randomized, controlled trial conducted in Japan that showed that edaravone slowed the rate of functional loss in ALS. Information on real-world effectiveness in patients treated with IV edaravone, however, is limited.

Objective:
To describe real-world treatment patterns in patients treated with IV edaravone collected by a provider of home and alternative-site infusions.

Methods:
A retrospective cohort study was conducted using the Soleo Health de-identified database. Soleo Health is a provider of home and alternative-site infusions and specialty pharmacy services in the United States and collects data on patients receiving IV edaravone. ALS patients receiving IV edaravone for ≥3 consecutive months between August 1, 2017, and March 31, 2020, were selected. Variables collected included demographic information, clinical characteristics, treatment outcomes, ALS Functional Rating Scale-Revised (ALSFRS-R) scores, and quality-of-life measures such as energy levels, sleep patterns, stress levels, mood, and overall health and wellness. All variables were assessed descriptively.

Results:
A total of 167 patients were included in the analysis. Mean age (±SD) was 60.4±11.3 years, 53.3% were male, 60.5% were enrolled in commercial insurance plans, and mean baseline ALSFRS-R was 37.1±9.24. During the 12-month follow-up period, 89 (53.3%) patients discontinued treatment. At the end of the study period, 78 (46.7%) patients continued treatment, with 64 (38.3%) having more than 12 months of continuous treatment with IV edaravone. The median treatment duration was 22.6 months (range, 12.2 to 30.8 months). In the patients who had more than 12 months of continuous IV edaravone treatment, the mean baseline ALSFRS-R score was 40.5±7.2 with a change per month in ALSFRS-R score of –0.63±0.54.

Discussion:
This analysis describes treatment persistence in a real-world setting. The change in ALSFRS-R score reported was consistent with changes observed in the pivotal Phase 3 study, Study 19, which showed a 33% slowing of the rate of functional decline with edaravone treatment and was the basis for IV edaravone's FDA approval. Future real-world analyses from other databases will examine time to real-world outcomes. It is hoped that this information will be useful to clinicians who prescribe IV edaravone for their patients with ALS.

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CMS-40: Trends in end-of-life practices in patients with ALS in the Netherlands from 1994 to 2016: a repeated survey cohort study

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Background:
After legalization in 2002, frequency of euthanasia in the Netherlands has increased from 2.4% in 1995 to 4.5% in 2015 (1). Between 1994 and 2005, euthanasia in ALS remained stable at 16-17% (2) and it is unclear whether it has followed the upward national trend.

Objectives:
To investigate the frequency and trends in physician reported End-Of-Life (EOL) practices in ALS in the Netherlands from 1994 to 2016 and to compare the perceived quality of end-of-life care between 2000-2005 and 2014-2016.

Methods:
A survey cohort study of patients diagnosed with ALS who died between 2014 and 2016. Physicians and informal caregivers were sent a questionnaire on end-of-life circumstances. End-of-life practices were classified based on physician-reported end-of-life decisions. Caregivers rated the patients' perception of different aspect of the quality of EOL-care during the last month before their death. Results were compared with those of two previous studies using the same questionnaire on patients who died during the periods 1994–1998 and 2000-2005. Chi square tests were used to determine trends in EOL practices and quality of care.

Results:
884 patients with ALS died between 2014 and 2016. 356 physician questionnaires were included (60% of respondents) and 211 caregiver questionnaires (38% of respondents). There was no difference in patient or disease characteristics between included and non-included patients.
Total EOL practices increased from 55-56% in 1994-2005 to 73% in 2014-2016 ($p < .001$), euthanasia increased from 16-17% to 39.6% ($p < .001$), forgoing life-prolonging treatment from 10-12% to 18% ($p < .05$), and continuous deep sedation from 15% to 22% ($p < .05$); intensified alleviation of symptoms decreased from 24-27% to 14% ($p < .001$).
Perceived quality of care in the 2014-2016 cohort was higher than in the cohort 2000-2005: sufficient information to ease suffering from 89% to 97% ($p < .01$); sufficient aids and appliances from 76% to 100% ($p < .001$); sufficient mental support from 78% to 95% ($p < .001$); sufficient symptom relieve from 75% to 87% ($p < .05$) sufficient knowledge and experience of healthcare professionals (HCP) from 66% to 91% ($p < .001$); sufficient confidence in HCPs from 68% to 95% ($p < .001$). Only adequate financial reimbursement decreased from 86% to 68% ($p < .001$).
Perceived quality of care in euthanasia did not differ from other EOL practices, no EOL practices, or unexpected sudden deaths.

Conclusions:
The increased frequency of euthanasia in ALS is in line with the upward national trend and is accompanied by an increase in perceived quality of EOL-care.

References:
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2. Maessen M, Veldink JH, Onwuteaka-Philipsen BD et al Neurology 2009; 73:954–61