THE RELATIONSHIPS BETWEEN FATIGUE, SLEEP AND DISABILITY IN MOTOR NEURONE DISEASE

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Keywords: fatigue, sleep, disability

Background: Fatigue is common to many neurological illnesses and is often rated as one of the most disabling symptoms. Fatigue seems to be prevalent in motor neuron disease (MND) but little is known about how it relates to sleep and other clinical features of the disease.

Objective: To assess the relationships between fatigue and sleep and demographic or clinical features of MND in a large population-based sample.

Method: The Neurological Fatigue Index for MND (NFI-MND) was administered to the patients with MND as part of the TONiC study, a multicentre, UK study of factors affecting quality of life in MND. Subjects were also asked to self-estimate duration of both day and night time sleep. Demographics and disease characteristics, including ALS Functional Rating Scale-revised (ALSFRS-r), were recorded by the clinical team. All scale scores were converted to interval level data by application of the Rasch measurement model. The results were analyzed using appropriate parametric tests.

Results: Four hundred and sixty-five records were available for analysis by March 2016. Mean age was 64.9 years, median disease duration 11 months, 60.6% had bulbar onset and the remainder, undetermined onset. Fatigue was greater in patients with muscle cramps, fasciculations and head drop (all p<0.001). Fatigue had a mild-to-moderate correlation with motor disability (p=0.49) but a non-meaningful correlation with respiratory impairment (p=0.28) as measured by the ALSFRS-R. 66% of patients slept in the day. There was seemingly no linear correlation between fatigue and duration of either day or night time sleep, but there was a striking V-shaped association of night sleep such that those that slept an average of 7 h at night had the lowest levels of fatigue.

Conclusion: Fatigue is largely independent of bulbar and respiratory involvement but generally increases with limb motor disability. Two thirds of patients sleep in the day; there is a V-shaped relationship between duration of nocturnal sleep and fatigue. Understanding these relationships might provide some insight into the underlying pathophysiology of fatigue in MND.

DOI: 10.1080/21678421.2016.1232068/001

ECONOMIC EVALUATIONS, COST STUDIES AND UTILITY STUDIES IN MOTOR NEURONE DISEASE/AMYOTROPHIC LATERAL SCLEROSIS: A SYSTEMATIC METHODOLOGICAL REVIEW

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Keywords: systematic review, economic evaluation, quality of life

Background: Although Motor Neurone Disease (MND/ALS) is a devastating condition which greatly affects patients’ quality of life, treatment options are very limited. With the present increase in research for treatments, future medications need a robust economic framework for assessment of their cost effectiveness. To date, there is no published systematic review of all published economic evaluations, cost studies and utility studies in MND/ALS.

Objectives: The aim of this research is to critique the methodology of current economic evidence relating to MND/ALS. These assessments will inform analysis of the health economics dataset being collected as part of the Trajectories of Outcomes in Neurological Conditions (TONiC) study in MND/ALS, which includes direct and indirect costs related to disease stage.

Method: Cost, economic and utility studies relevant to MND/ALS, written in English and published up until 2016, were identified from Medline, EMBASE, Econlit, NHS Economic Evaluation Database, the Health Economics Evaluation Database and reviewed critically for their methods.

Results: Sixteen economic evaluations, 22 cost studies and three utility studies were identified. Most economic studies were model based (n=13) with the majority of these studies focusing on riluzole (n=9). Economic studies adopted a health services costing perspective (n=11), a societal perspective (n=1) or both (n=4). Half of the economic evaluations were conducted in the UK (n=8). Outcomes in economic studies included the reporting of Incremental
Cost-effectiveness Ratios (ICERs) (n=6), Quality Adjusted Life Years (QALYs) (n=3), or both QALYs and ICERs (n=6). Cost studies in this review all considered direct medical costs (n=22), with half including an indirect costs component to their analysis (n=11). Cost studies were American (n=11) or European (n=9) with two from Asia. Few cost studies reported disease stage specific costs (n=3). Utility studies used the EQ-5D (n=2) and the standard Gamble (n=1) to elicit patient preferences for disease states. The methods adopted by the included papers in this review are analyzed in our study.

Discussion and conclusion: Economic evaluations in MND/ALS suffer from significant methodological issues, such as a lack of relevant data, limited scope and significant uncertainty with the disease course. Cost studies were hampered by high variability and country specific characteristics such as differences in healthcare systems and delivery of care. The utility studies presented in this review notably suffered from concerns over the appropriateness of generic utility measures, potentially unrepresentative cohorts and a lack of consideration for the impact of treatment upon caregivers. Our analysis shows a clear need for up to date and robust data for future unbiased assessment of the cost-effectiveness of interventions in MND.

Acknowledgements: The authors would like to thank the Motor Neuron Disease Association UK for their funding.

DOI: 10.1080/21678421.2016.1232068/002

P383 THE PATIENT JOURNEY TO A NATIONAL ALS CLINIC: DELAYED DIAGNOSIS AND ECONOMIC COST

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Keywords: referrals, interventions, cost

Background: There are currently no effective disease modifying therapies for Amyotrophic Lateral Sclerosis (ALS) and management is symptomatic. A diagnosis of ALS is primarily based on the physician’s interpretation of clinical symptoms and signs, and investigations to exclude other causes, and this may prolong patients’ journey to multidisciplinary care. Earlier diagnosis can enable more effective symptom management care planning and can alleviate caregiver burden.

Methods: We have detailed the journey of a national cohort of ALS patients (n=156) from the time of first symptom to presentation at the ALS multidisciplinary clinic (MDC), using data from a detailed chart review and national register. Key milestones in the patients’ journey to the ALS clinic were analyzed, including pattern and frequency of consultations, clinical interventions, and associated economic cost.

Results: A majority of patients was male (60%), 65 years of age and over (54%), 72% were of spinal onset. Twenty seven patients received an alternative diagnosis prior to attending the clinic. Time from onset of first symptoms to ALS diagnosis was a mean of 15.1 months (median, 11). There was a mean interval of 17.4 months (median 12.5) from first symptoms to arrival at the MDC. There was a mean of 3.23 (median, 3) consultations with health care professionals prior to MDC attendance, most commonly neurologists and General Practitioners. Direct referral by a General Practitioner to a neurologist reduced diagnostic delay. Bulbar ALS was associated with shorter time from symptom onset to diagnosis, and age was weakly and positively predictive of cost prior to the ALS clinic attendance. Mean cost prior to attending the MDC was €2,392 per patient. We estimate that expedited referral to the ALS clinic could have reduced costs by an estimated €901 per patient.

Conclusion: Development of a standardized pathway with early referral to neurology of patients with suspected symptoms of ALS can reduce diagnostic delay and limit unnecessary investigations. This study suggests that a reduction in healthcare spending is possible from improvement in referral pathways to the ALS clinic.

Acknowledgements: The project is funded by the Health Research Board (HRB) Dublin as part of the HRB Interdisciplinary Capacity Enhancement Awards.

DOI: 10.1080/21678421.2016.1232068/003

P384 PERSONNEL COSTS OF A MULTIDISCIPLINARY ALS CLINIC

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Keywords: cost, multidisciplinary care

Background: People with ALS who receive their care by experienced multidisciplinary teams live longer and have improved quality of life (1). Costs of running an ALS multidisciplinary clinic range from $258 to $806 per patient per clinic across centers (2). In the United States, most insurance reimbursement only provides consultation and follow up coverage for providers (physicians, nurse practitioners, physicians assistants) leaving personnel cost for the multidisciplinary care team to be covered through other means; foundation support and/or philanthropy.

Objective: To examine the personnel cost of an ALS multidisciplinary clinic that is not reimbursed by health insurance in the United States.

Methods: Reviewed 2015 annual budgeted personnel costs (salary, fringe and overhead) to adequately care for 409 unique ALS patients in 1285 ambulatory office patient encounters, including 300 new patient encounters and 985 follow-up encounters. This budget identified percent of efforts of the multidisciplinary care team, including: 0.58 physician, 0.3 nurse practitioner, 3.6 registered nurse, 1.55 physical therapist, 0.2 speech and
language therapist, 1.5 resource specialist, 0.05 practice manager, 2 patient coordinators. We calculated the total insurance reimbursement revenue for billable physician or nurse practitioner encounters over the same time period.

**Results:** Annual personnel costs to support 1285 patient encounters with allocated staff in an ALS multidisciplinary clinic is $977,808 with an average cost per patient per clinic of $761. Insurance reimbursement totalled $338,398 ($263 per patient per clinic) leaving a shortfall of $639,419 ($497 per patient per clinic).

**Discussion and conclusion:** Only one-third of the personnel costs to provide optimal care for people with ALS in a multidisciplinary clinic are covered by insurance reimbursements in the United States. While the remaining coordination of care are not billable; in between visit phone triage, medications refills, durable medical equipment requests, letters of need, disability paperwork and prior authorizations. Foundation and philanthropy support are the primary resources necessary to support non-provider salary support as there are few clinical support staff grants available.

Further discussion and research is needed to determine if the same optimal care and improved quality of life is achieved through non-provider dominant care.

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DOI: 10.1080/21678421.2016.1232068/004

**P385 EMOTIONAL EXPERIENCE IN PATIENTS WITH AMYOTROPIC LATERAL SCLEROSIS WITHDRAWING FROM LONG-TERM VENTILATION**

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**Keywords:** palliation, end of life, termination of ventilation

**Background:** Non-invasive ventilation or tracheotomy with invasive ventilation are treatment options in ALS. However, a proportion of patients receiving long-term ventilation (LTV) decide to have it withdrawn.

**Objectives:** The objective of this study was to analyze the emotional experience of ALS patients withdrawing of long-term ventilation (WLTV).

**Method and sample size:** The study aimed to explore the emotional impact of the decision for WLTV in patients with ALS, their relatives and professional care givers. In a systematic survey, patients (n=8) were prospectively studied before receiving WLTV. Additionally, 20 relatives of patients with ALS and WLTV were surveyed retrospectively. The questionnaire consisted of 20 items (23 items for relatives). 12 items adopted to were adapted to the a visual analogue scale (VAS) with, an additional 8 open questions plus 3 open questions for relatives.

**Results:** ALS patients showed lower satisfaction with LTV (VAS 3.6/10, n=5) as compared to their relatives (VAS 6.6; n=14). Latency between decision-making and realization of WLTV was 5.3 months. Patients informed their families about the decision of WLTV at different times: >12 month (n=4), >3 month (n=8); >1 month (n=4); <1 week (n=2). The patient’s wish for WLTV was related to loss of communication (66%), followed by loss of mobility (44%) and hopelessness for cure (32%). The patient’s option to determine his or her date of death by means of WLTV was experienced as a relief rather than a burden by all patients (10/10; n=6). However, emotions of family members were dominated by sadness (8.6/10) and the loss of a loved-one (8.3/10). Wishes of patients were more strongly backed by relatives (VSA 8.9/10) than by care providers (5.4/10)

**Conclusion:** Patients evaluate LTV less positively than their relatives. Decision-making of WLTV is a process of several months. In general, patients experience their decision of WLTV as an emotional relief. Family members supported the patient decision of WLTV. In contrast, patients experience the attitude of professional care providers as less supportive. Our results demonstrate different attitudes towards LTV among patients, relatives and professional care givers.

DOI: 10.1080/21678421.2016.1232068/005

**P386 DETERMINING THE IMPACT OF A DESIGNATED MULTIDISCIPLINARY CARE CENTRE IN CAMBRIDGE ON THE NATURAL HISTORY AND MANAGEMENT OF MOTOR NEURONE DISEASE**

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**Keywords:** multidisciplinary care, survival, NIV

**Background:** The impact of specialist care in the management of MND has been a focus of longstanding interest. In the UK, the multidisciplinary team (MDT) approach has been consolidated by the establishment of designated MND Care Centers, although data regarding their impact on care are limited. The Cambridge MND Care Centre was established in 2005 and serves the populations of Cambridgeshire and the surrounding counties of eastern England.

**Objectives:** We set out to investigate the impact of the Cambridge MND Care Centre on the care provided to patients before and after its establishment in 2005.

**Methods:** We undertook a retrospective medical record-based study. Two cohorts of patients given a diagnosis of MND were identified: 74 patients were identified who
attended Cambridge between 2001 and 2003 and 144 patients attending between 2011 and 2013. Demographic and clinical data were collected and a comparative analysis of the two cohorts performed.

**Results:** No significant difference was seen on survival curves where all patients from both cohorts were included. However, a trend for increased survival could be observed after 2 years in the post-care centre cohort when patients with disease duration longer than 20 years were excluded. A significantly greater proportion of patients were referred to the regional respiratory service in the post-care centre cohort for consideration of non-invasive ventilation (NIV) (74.3% vs. 63.5%, \( p<0.05 \)), although there was no significant difference in the proportion of referred patients offered the treatment.

**Discussion:** Our data suggest that by consolidating the care of patients in a specialist environment after 2005, the proportion of patients living with MND in our region who were referred for NIV increased significantly. We believe that this is likely to be a surrogate marker of a more structured approach to care, as domiciliary NIV was being offered by our regional respiratory centre before becoming standard practice in MND, making referral the limiting factor. Although no significantly increased survival was seen following the establishment of the care centre in 2005, there are suggestive data pointing towards beneficial changes 2 years from symptom onset which are unlikely to be attributable to the use of NIV alone.

**Conclusion:** Our study therefore adds further evidence concerning the impact of the establishment of a designated multidisciplinary care centre on the management of patients living with MND, while also highlighting the local and general difficulties associated with such retrospective clinical studies in such a clinically heterogeneous disorder.

**Acknowledgements:** MND Association.

DOI: 10.1080/21678421.2016.1232068/006

**P387 MOTOR NEURONE DISEASE: STAFF PERSPECTIVES OF THE GOAL SETTING PROCESS WITHIN A COMMUNITY MULTIDISCIPLINARY TEAM**

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Keywords: goal-setting, community multidisciplinary care, quality of life

**Background:** Goal-setting within neurological rehabilitation teams is a fundamental and widely practiced process. Little is known about the goal-setting process in palliative conditions such as Motor Neuron Disease (MND). MND is a rapidly progressive condition which is unpredictable in nature. The management of clients with MND is a challenge with intervention mainly aimed at supporting decline in function and decision making for inevitable future changes. Involving clients in the goal-setting process is essential in supporting quality of life; however the mechanism by which this is best done is unclear (1,2).

**Objective:** The aim of this project was to explore the perspectives of health professionals working in a community setting, who goal set with patients with a diagnosis of MND; considering the reasons for, and the perceived facilitators and barriers to goal-setting for this client group.

**Methods:** A qualitative enquiry was employed in this study with the use of focus groups (FGs) and the student as a research tool to collect data. A total of 17 participants were recruited into the study forming 3 FGs. FGs were audio recorded, transcribed and analyzed using thematic analysis.

**Results:** A total of 8-themes were established, these were; fostering hope, therapists holding the goal-setting power, nurturing the therapeutic relationship, patient acceptance and adjustment, managing change and uncertainty, professional models and practices, goal-setting and educational conventions and future changes.

**Discussion and conclusions:** Participants from this study were found to value the use of a patient-centered approach to goal-setting, however goal-setting was discussed as being a predominantly problem focused and therapist led process. A number of influencing factors were established, such as the responsibility to sustain hope and the implicit need to support change and minimize the related risks, by the health professional. The study also highlighted the paradox that exists with managing palliative conditions such as MND within a rehabilitative context, finding physiotherapists as a professional group to struggle the most when managing this. This study concludes that further consideration and action needs to be given by health services in overcoming factors which influence the patient-centeredness of goal-setting, supporting the way to a truly patient-centered health service.

**Acknowledgements:** This project was completed as part of a Master of Science in Neurorehabilitation, I would like to thank Dr Cherry Kilbride from Brunel University London for her support and encouragement. I would also like to thank the Royal Free Neurological Rehabilitation Centre for supporting the study.

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DOI: 10.1080/21678421.2016.1232068/007

**P388 ALS MEDICAL NEEDS AND THE REGIONAL MEDICAL RESOURCE SURVEY IN JAPAN**

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**Background:** There are many factors to consider in the management of ALS: ensuring maintenance of the medical treatment environment and informed caregivers, ensuring the long-term care destination and short-term inpatient facilities, providing psychological support, etc. In Japan, by the arrangement of the establishment and specialists of the incurable disease medical care network of each prefecture, a relatively smooth care pathway has been provided in some areas, but we think that in many areas it is still inadequate.

**Objectives:** To understand the medical resources provided, by seeking opinions about the incurable disease medical specialists from ALS patients and their families, as the disease progresses, by comparing the current state of the health care resources of each region, taking into account regional characteristics.

**Methods:** 1. Target: ALS patients and their families; 2. Survey method: anonymous self-administered questionnaire; 3. Survey items: recuperation status after diagnosis, knowledge and use of state or public support systems, source of information about the disease (such as the presence or absence of psychological support), multiple-choice questions that ask about medical resources and their level of satisfaction; 4. Recruitment methods: fundamental research team members in neurodegenerative disease area from incurable diseases health professionals who obtained the consent and mailing lists, were asked to distribute the questionnaire; 5. Ethical considerations: approved by the Kyushu University physician member association for people with neuromuscular diseases, funded this study.

**Results:** The survey was completed by 151 people. The average age was 63.3 years of age, 64.9% were men. Of the respondents, 111 patients (73.5%) were fitted with invasive/non-invasive ventilator. Regional distribution of respondents, Hokkaido and Tohoku 3.3%, Central 12.5%, Kanto13.9%, Kansai 17.2%, China 12.5%, Shikoku 2.6%, was Kyushu 37.7%. 16.5% of respondents continue to see incurable disease medical specialists, 21.1% had received/experienced a consultation. Respondents commented that there wasn’t enough nationwide awareness yet. In terms of the nature of the consultation, “consultation of home care,” was received in 21% of people, “system provides information of” in 18%, “information provided about assistive devices” in 15%, “consultation of hospitalization, hospital change” in 14% and “disease and provide information about the treatment” was 14%.

**Conclusions:** Depending on the location and interventions of the timing of the incurable disease medical specialists, it was thought that the ease and consultation support of the contents of the access is different.

**Keywords:** the public support systems, home care, medical needs

**P389 HOW 95 PERCENT OF ALL DANISH ALS PATIENTS USE A NATIONAL CENTRE OF REHABILITATION EXPERTISE**

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**Keywords:** multidisciplinary care, co-ordinating interface, epidemiology

**Background:** Ninety-five percent of all people diagnosed with ALS in Denmark accept referral from the hospital to the national Rehabilitation Centre for Neuromuscular Diseases (RCND). This centre develops and supports multidisciplinary approaches in rehabilitation and palliation, both at a personal, family and community level, in line with recommendations to establish a co-ordinating interface between neurology, rehabilitation and palliative care (1). The individual use of specific RCND services and interventions in relation to disease progression and other key variables has not been studied systematically.

**Objectives:** To describe, quantify, and assess the use of 11 key RCND services and interventions in relation to age, sex, disease progression, geography, marital status, and membership of the disability association for people with ALS.

**Methods:** A descriptive statistical 4 year follow-up profile of the individual use of RCND services and interventions in a total one calendar year cohort of patients with ALS referred to RCND.

**Results:** All patients with ALS (N=135) referred to RCND in 2011 was identified. The total individual use of 11 services and interventions in the period 1 January 2011 to 31 December 2014 was retrieved from RCND records. The individual use was calculated in relation to age, sex, disease progression, geography, marital status, and membership of the disability association for people with ALS. Patterns of individual use mirrored variability in relation to age and disease progression, but also proved earlier assumptions to be wrong, for example regarding the actual proportion of patients participating in peer group courses which turned out to be about 50 percent of eligible patients (2).

**Discussion and conclusions:** On the one hand, 4-years follow-up profiles of use of services and interventions confirmed that the population of patients with ALS consists of subgroups with well-known needs for support and rehabilitation. On the other hand, the profiling survey applied in this study revealed some surprising correlations, for example between disease progression, marital status, and participation in peer group courses. The study underlined that complexity in each case is a standard challenge in ALS rehabilitation. It means that standardization of procedures, interventions, and programs is limited. Pre-admission assessment to services and interventions must carefully take into account the individual complexity. Similar patients with ALS need different rehabilitation.

**Acknowledgements:** Muskelsvindfonden, the Danish member association for people with neuromuscular diseases, funded this study.

**Keywords:** multidisciplinary care, co-ordinating interface, epidemiology

**DOI:** 10.1080/21678421.2016.1232068/008
Patients with bulbar onset showed a worsening in the fields of speech and swallowing and the patients with appendicular presented worsening in the fields of mobility of upper and lower limbs. Issues related to communication and the emotional aspects interfere in a similar way, in the QL of both groups.

Background: Few studies have examined an ALS patient’s attitude towards life-prolonging measures, such as NIV, gastrostomy, or tracheostomy, and no data is available regarding how the attitude of a patient towards various interventions might evolve or change during the course of disease progression (1,2). The multidisciplinary ALS clinic at the Montreal Neurological Institute and Hospital currently raises the topic of life-prolonging measures at multiple instances throughout the course of disease progression, with the hypothesis that a negative attitude might become more positive as a patient’s disease progresses. The goal of this study is to either support this hypothesis, or provide new insight into how and when these discussions should be approached.

Objective: The purpose of this study is to identify an ALS patient’s attitude towards life-prolonging measures during various stages of disease progression, and to evaluate the current practices in the multidisciplinary ALS clinic.

Methods: This study has a cross-sectional, observational design. Basic demographic data, as well as ALSFRS-R scores, were collected. Patients expressed their attitudes towards various life-prolonging measures on a pre-set scale, and were then encouraged to elaborate in open-ended interviews. Patients having already received an intervention were excluded from this study.

Results: Data from twenty-eight (n=28) ALS patients was analyzed. Patients demonstrated a more positive attitude towards life-prolonging measures in general as the disease progressed; however, attitudes towards both gastrostomy and tracheostomy remained negative until the end stage of the disease. Patients having participated in, or currently involved in, a clinical trial were more in favor of discussing tracheostomy ($p<0.05$). Qualitative analysis of the open-ended discussions revealed multiple themes that contribute to a patient’s decision regarding life-prolonging measures. Furthermore, use of interventions in the clinic were evaluated for the last 15 years, and there is an increasing trend in both BiPAP initiation and...
PEG placement. Initiation of PAV remains constantly sparse over time.

**Conclusion:** Our study supported the hypothesis that patients develop a more positive attitude towards life-prolonging measures as the disease progresses. Our results also support the multidisciplinary ALS clinic’s current practices of raising the topic of interventions at multiple instances during disease progression. Finally, we identified key themes that contribute a patient’s decision regarding interventions, which will contribute to future discussions undertaken by the clinic staff.

**Acknowledgements:** We would like to thank the ALS patients that participated in this study, as well as the Montreal Neurological Institute & Hospital’s multidisciplinary ALS clinic for their support.

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DOI: 10.1080/21678421.2016.1232068/0011

**P392 PRACTICE PATTERN OF NON-INVASIVE VENTILATION AND ITS IMPACT ON END-OF-LIFE CARE IN PATIENTS WITH AMYOTROPHIC LATERAL SCLEROSIS AMONG CANADIAN CARE PROVIDERS**

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**Keywords:** non-invasive ventilation, end-of-life

**Purpose:** Over two stages, the study will (1) describe current non-invasive ventilation (NIV) usage patterns amongst Canadian ALS healthcare providers; (2) compare/contrast with previous practice patterns (1); (3) explore barriers to NIV access, and similarly; (4) identify the impact and challenges of NIV on end-of-life care in ALS.

**Methods:** In both stages, healthcare professionals (ie physicians, respiratory therapists and nurses) at major Canadian ALS care centers were sent a web-based survey. In the second stage, palliative care physicians were also invited to participate. The first stage of the study explored NIV usage patterns. Participants were asked to provide input on practice demographics, access and initiation of NIV and follow-up of NIV. The second stage of the study explored the impact of NIV on end-of-life care in ALS. Participants were asked to provide input on practice demographics and how they carry out palliative care, end-of-life discussions and NIV withdrawal in ALS patients using NIV. Quantitative data were analyzed with descriptive and comparative statistics, while qualitative data were analyzed using interpretative phenomenological analysis method to identify emergent themes. When possible, the data was compared with previous findings.

**Results:** 26 participants responded in the first stage. Median NIV usage was 39% (range 10–100%), about double of what was previously reported (18%). Mean wait times from referral to routine and urgent NIV initiation were 13 (95% CI 9–17) and 5 (95% CI 3–7) days, respectively. NIV was most commonly initiated in clinic (68%), while 38% report having access to home-NIV initiation. Compared to previous findings, clinicians are more reliant on pulmonary function measures than blood gases for deciding NIV initiation. In terms of barriers, lack of social support (62%) and cognitive impairment (46%) were the most common deterrents to initiating NIV. Barriers to access can be stratified to patient, clinical, institutional and regional levels. The second stage of the study is nearing completion. We hypothesize that challenges with respect to NIV withdrawal would include a lack of a standardized approach and a lack of clinical and emotional support for the staff involved in the withdrawal process.

**Conclusions:** Compared to previous findings, NIV usage in Canada is more prevalent, and its initiation sooner and easier. Management of NIV intolerance may also have improved. Clinicians are more reliant on pulmonary function measures than blood gases for NIV initiation. Despite improved timeliness and access to NIV, barriers are still present; however, their identification will aid policy makers in improving timely and efficient access to NIV.

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DOI: 10.1080/21678421.2016.1232068/0012

**P393 RE-EXAMINING THE UTILITY VALUE OF MECHANICAL VENTILATORS TO ENABLE THE LONG-TERM SURVIVAL OF ALS PATIENTS**

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**Keywords:** mechanical ventilators, independent living, outside care providers

**Background:** The use of mechanical ventilators by ALS patients allows for their long-term survival; however, the rate of use is less than 30% in Japan, and extremely low, at approximately 3%, in Europe and North America. Japanese ALS patients have strongly criticized the social tendency to deny the use of mechanical ventilators to extend life.

**Objectives and methods:** The objectives of this report are to interrogate and examine the ALS patient’s decision-making process about the utility value for mechanical ventilation: how this is perceived for ALS patients, how society might be made to perceive it, and the potential for offering assistance for it. The subject of this investigation
is researcher observation of the lives of ALS patients as well as their testimonies. Specifically, the author focuses on the decision-making process in the choice to use mechanical ventilation, and demonstrates the continuing stigmatization of mechanical ventilators from the feelings of discord observed in patients regarding its use.

Results: It becomes clear that ALS patients have a negative perception towards the use of mechanical ventilation owing to the facts that its use inhibits their ability to leave the house and communicate freely, and that there is an intense burden of care placed on family members. On the other hand, in Japan, ALS patients who use mechanical ventilation and are nearly paralyzed utilize systems of 24-h care by outside care providers, and live their lives holding regional jobs. It was discovered from their testimonies that they have a positive perception towards the potential for a longer life through the use of mechanical ventilation.

Discussion: The utility value of mechanical ventilation is in its potential to offer long-term survival of patients. The results of this investigation reveal that the value of mechanical ventilation is evaluated negatively due to the impasses of family care and the problems that emerge through long-term survival. The negative evaluation of the long-term survival of ALS patients through mechanical ventilation by social tendencies suggests that the survival of ALS patients in society through the use of mechanical ventilation is relegated to a lower priority.

However, advocacy by ALS patients groups and patients who actually live using mechanical ventilation demonstrates the potential for life extension through mechanical ventilation, and the perception of such is prompting a change in the decision-making process regarding the use of mechanical ventilation. The lifestyles of these patients and the advocacy of such, commonly champions the enrichment of social systems. Through the distribution of the burden of family care and the offering of assistance in communication and trips outside the house, the utility value of mechanical ventilation in extending life is made visible, the survival of ALS patients through the use of mechanical ventilation is affirmed.

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**P395 HOW CAN WE EDUCATE ABOUT PALLIATIVE CARE FOR ALS?**

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Keywords: education, palliative care, end of life

**Background:** Palliative care for ALS, which includes the early stages of the disease to the end of life and grief care, require not only medical knowledge but also ethical consideration. Ethical issues have more than one answer, and it is difficult to get good evidence and to educate efficiently. In Japan, palliative care was developed mainly for pain management of cancer patients. Likewise, education courses on palliative care provided by the government are also for cancer patients. Palliative care for non-cancer patients has just started, and many doctors have little experience. Thus education in palliative care for non-cancer patients is very important. Due to this, we have been organizing an education course of palliative care for ALS patients once a year since 2011. The program includes: patient-doctor relationships, decision making for PEG, decision making for respirators, support for decision making, ethical issues for the end of life by ethics experts, legal issues for the end of life by lawyers, how to use opioids for ALS patients, and a case study of palliative care for neurodegenerative diseases. The program includes: mixed small group discussion (SGD), role playing (RP), and lectures.

**Objective:** To evaluate the efficacy of this program for palliative care in ALS.

**Method:** We conducted the survey where the participants answered a questionnaire (a) right after the end of the program, and in March 2016 we sent another questionnaire (b).

**Results:** (a) The survey right after the end of program (Five ratings of evaluations: 1 extremely positive, 2 considerably positive, 3 positive, 4 considerably negative, 5 negative): the value of this program: 1–58%, 2–39%; the need of education like this program: 1–82%, 2–10%; should it continue in future?: 1–82%, 2–17% b) the survey at 6 months to 5 years after the program (anonymity-style questionnaire, return rates was 40%): how was this program helpful for your clinic: extremely helpful 45.2%, considerably helpful 50.7%, not very helpful 4.1% (the reason was they do not have ALS patients), not helpful 0%. The need of education like this program: 1–82%, 2–17%. Should this program continue in the future?: 1–76%, 2–21%. According to this result, this program was shown to be very useful even after many years. They highly valued this education method, such as discussion of problems in SGD, exploring by experiencing RP and finally having answers to questions in lectures.

**Conclusion:** There are few opportunities for palliative care education for ALS in Japan, and the empowering workshop like the one reported in this study is recommended and effective for clinical practice.

DOI: 10.1080/21678421.2016.1232068/0015

**P396 THE PALLIATIVE CARE NEEDS OF PEOPLE WITH ADVANCING NEUROLOGICAL DISEASE IN IRELAND**

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Keywords: palliative care, neurology

**Background:** Within Ireland, there is a lack of consensus and direction with regard to the palliative care needs of people with advancing neurological disease. Staff members and volunteers of organizations that are members of a neurological umbrella organization informed this study. The population total was fourteen.

**Objectives:** 1. Explore the palliative care needs of people with advancing neurological conditions from the perspective of Neurological Alliance of Ireland member organizations to include the Irish Motor Neuron Disease Association; 2. Identify steps required to enhance palliative care for people with advancing neurological illnesses in Ireland.

**Methods:** A literature review and a series of qualitative interviews were carried out. Interviews were recorded and analyzed using NVivo to identify themes.

**Results:** 1. The literature review and interviews identified that ambiguity exists around the terminology of palliative care. Palliative care is primarily associated with specialist services; 2. The following issues arise for organisations: Difficulty accessing palliative services; Uncertainty when palliative care begins; Planning Ahead; The requirement for training; 3. The following themes were identified by participants as a means of enhancing palliative care for this group: Greater understanding of symptoms and unpredictable nature of illnesses; Emphasis on interventions that support quality of life and planning ahead; Availability of palliative care approach earlier; Enhanced multidisciplinary and psychosocial care; Increased equitable access to specialist palliative care; More training for staff.

**Conclusion:** Collaboration among all stakeholders is required to ensure the palliative care needs of those with advancing neurological disease are adequately addressed.

**Acknowledgements:** This work, commissioned by the Irish Hospice Foundation and Neurological Alliance of Ireland, was carried out by Dr John Weafer. The work was guided and supported by a working group made up of members of Neurological Alliance of Ireland. Representatives from the following organisations took part in this work: Irish Motor Neuron Disease Association; Brain Tumor Ireland; Cheshire Ireland;
Huntington Disease Association of Ireland; Multiple Sclerosis Ireland; Parkinson’s Association of Ireland; Progressive Supranuclear Palsy Association (PSPA) Ireland

DOI: 10.1080/21678421.2016.1232068/0016

P397 PALLIATIVE CARE OUTCOMES IN ALS
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Keywords: ALS, palliative care outcome scale

Background: Studies have shown that early palliative care intervention from time of diagnosis of Amyotrophic Lateral Sclerosis (ALS) is beneficial to patients. Palliative care in Ireland is moving from a “cancer only” discipline to become an integral part of the management of non-malignant conditions such as ALS and other chronic diseases such as COPD and heart failure. Palliative care intervention has been shown to improve quality of life and also aid in symptom control. The Palliative Outcome Scale (POS) is an outcome measurement tool which is used to assess patients’ change in health over time. Evaluation of these outcomes can help identify the triggers for palliative care interventions in ALS.

Objective: The aim of this study was to evaluate whether the setting of assessment (POS recorded at home or in clinic) influenced palliative care outcome scores. The project also aimed to assess whether variables such as ALS subtype and gender influenced POS score.

Methods: A prospective study, in which POS questionnaires were administered to 100 ALS patients who attended the National ALS clinic in Beaumont Hospital were analyzed. Data was collected in two settings at three time point intervals, either in the patients home by a neuropsychology researcher or in the outpatient department (OPD) by a senior neurology registrar in Beaumont Hospital, Dublin.

Results: Home assessed POS scores were notably higher than clinic assessed (p> 0.001). No significant differences were found on analysis of ALS subtype or gender. A positive correlation was found between POS scores and disease progression.

Conclusion: Inclusion of POS into the matrix of tools used for palliative care assessment in ALS can prove useful in highlighting changes in patients’ needs and essentially triggers for care interventions.

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DOI: 10.1080/21678421.2016.1232068/0017

P398 COLLABORATION BETWEEN ALS AND PALLIATIVE SPECIALISTS IN DENMARK
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Keywords: palliative rehabilitation, collaboration

Background: Throughout their entire disease course, persons with ALS get support from interdisciplinary ALS teams at neurology and pulmonary departments, and the Danish Rehabilitation Centre for Neuromuscular Diseases (RCFM). In addition, they receive day-to-day care and treatment provided by local home care teams, possibly a team of non-professional helpers, and their general practitioner. In some cases, when the disease reaches a more advanced stage, the need for relief increases and more intensive, palliative interventions are required, sometimes in a setting other than the home or neurology department.

Organization of palliative care: In their recommendations for palliative care (2011), the Danish Health Authority writes that the intervention should be organized at the basic level in the primary sector by home care, the GP and the local hospital and at specialist level by a hospice and an outgoing palliative team. By incorporating palliative care as best practice in ALS rehabilitation and work routines, assisted by palliative specialists if needed, we can ensure that persons with ALS and their families and helpers receive better palliation.

Objectives: To establish and develop formalized partnerships between ALS and palliative specialists, and to develop a set of evidence-based guidelines to improve the palliative effort in ALS rehabilitation.

Methods: At the beginning of 2014, RCFM presented the project to nine palliative specialist units in Eastern Denmark and informed them about the disease and the organization of ALS care and treatment. At the beginning of 2015 and 2016, meetings were held at each of the nine units where shared cases were discussed and the project evaluated. A joint project meeting with the nine specialist units, four ALS teams, the respiratory department at Righospitalet, and RCFM was held in September 2015 where specific problems and the future of the project were discussed. Another meeting is scheduled for September 2016.

Results: The project has resulted in closer collaboration on shared ALS patients between palliative specialists. Two out of 62 patients who died in 2011 received help from palliative specialists. During the project period 2014–2015, the number of patients seeking help from palliative specialists had increased to 25 of which 18 out of 117 died with support from palliative specialists.
Conclusion and recommendations: Joint pre-referral visits; Collaboration on shared ALS patients; Design of an ALS data card for hospice use; Personal contact to collaborators strengthens the collaboration.

DOI: 10.1080/21678421.2016.1232068/0018

P399 WHAT ARE JAPANESE NEUROLOGISTS’ OPINION ON MORPHONE/SEDATIVE USE IN ALS PATIENTS AT THE END OF LIFE STAGE?

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Keywords: morphine, sedatives, palliative care

Background: In Japan we have had problems in palliative care for ALS patients, for example morphine was not approved for use by national health insurance. When it became approved, it was then recommended in the revised Japanese ALS guideline, published in 2013. But, has Japanese neurologists’ attitude towards use changed?

Objectives: To clarify the current Japanese neurologists’ opinion on morphine/ sedatives use in palliative care for ALS patients and its change since the first nationwide survey.

Methods: We performed a nationwide survey about palliative care in ALS patients. We sent a questionnaire to 5144 board certified neurologists.

Results: 1470 neurologists responded (response rate 27%). Q2-27: (1) Should be positively used (2009: 23.6%, 2012: 25.8%, 2015: 23.1%); (2) Not willing to use but unavoidable (2009: 51.6%, 2012: 55.2%, 2015: 56.2%); (3) Must not be used (2009: 4.6%, 2012: 4.6%, 2015: 4.5%); (4) Cannot decide (2009: 17%, 2012: 14.4%, 2015: 12.8%); (5) Others (2009: 5.6%, 2012: 2.6%, 2015: 4.6%). Neurologists who have prescribed morphine to more than four patients answered (1) (2012: 21.1%, 2015: 21.8%) (2) (2012: 62.7%, 2015: 51.7%) (3) (2012: 1.4%, 2015: 9.6%) (4) (2012: 13.4%, 2015: 14.4%) (5) (2012: 0.7%, 2015: 1.7%). Q2-28: (1) Identify with Euthanasia (2009: 3.6%, 2012: 1.9%, 2015: 2.9%); (2) different from euthanasia but with the identical effect (2009: 24.6%, 2012: 21.9%, 2015: 22.8%); (3) Totally different from Euthanasia (2009: 66.6%, 2012: 71%, 2015: 69.8%); (4) Others (2009: 7.1%, 2012: 5.2%, 2015: 4.5%).

Discussion: Morphine became approved for dyspnea in ALS patients and the revised Japanese ALS guideline published in 2013 recommends its use. But Japanese neurologists’ opinion has not changed much. It is interesting that for neurologists, who have prescribed morphine to more than four ALS patients, the rate who answered “sedatives must not be used” has increased (1.4—9.6%). We assume that appropriate use of morphine made them reluctant to use sedatives which have a higher risk of respiratory distress.

DOI: 10.1080/21678421.2016.1232068/0019

P400 PROVIDER ORDERS FOR LIFE SUSTAINING TREATMENT (POLST)-A TOOL FOR DOCUMENTING CHOICES OF ALS PATIENTS

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Keywords: advance directives, end of life choices, shared decision-making

Background: Provider Orders for Life Sustaining Treatment (POLST) are a set of portable medical orders that offer the ability to limit interventions (do-not-resuscitate or allow natural death). They do not replace advance directives. The form is completed as a result of shared-decision making in which the patient discusses their values, beliefs and goals and the provider presents diagnosis, prognosis, alternatives including the benefits and burdens of life-sustaining treatments.

Objective: Our goal was to analyze patient characteristics and the shared-decision making pattern captured through POLST at our clinical center.

Method: Two clinical datasets about cognitive and gastrostomy placement were used to review information regarding POLST. Logistical regression was used in the analysis.

Results: Eighty-two patients were studied. 50% were female with average age of onset of 56 years. 32% were bulbar. 10% had familial ALS. 61% of patients had a POLST order on file. 102 POLST forms were available with 35 patients having a single POLST document on file. 15 patients had at least two POLST orders during the course of their illness. 32 did not have a form. 57 of the forms were filled out to be DNR (56%) with 22 specifying comfort care (39%). 35 forms indicated limited interventions (61%). 12 documents (12%) were checked as full code with 7 opting for limited interventions (58%). Of the patients who had multiple forms, 11 patients showed expected progression in choices (from full to limited to comfort care). Only one patient had reversal to a more aggressive form of intervention (from comfort to limited intervention) two months after feeding tube placement was performed. Those using non-invasive ventilator support were more likely to have a POLST (76 vs. 33%,
There may have been a slight bias towards comfort or limited interventions in female patients as compared to male patients (38% vs. 12%, $p=0.1$). Patients older than 60 years of age were more likely to choose comfort measures only (35% vs. 22%, $p=0.35$). There was no association with the rate of decline of ALSFRS, marital status or with having young children under age 18 years old in the household.

**Discussion and conclusion:** POLST appears to be a viable tool for shared-decision making in patients with ALS who wish to limit interventions. A majority of patients at our center had a POLST in place. The choices were in keeping with an expected shift in values and goals in the course of disease progression. Systematic capture of patient and caregiver satisfaction and assessment of decisional conflict would be important next steps in the assessment of POLST as an effective decision aid.

DOI: 10.1080/21678421.2016.1232068/0020