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# THEME 9 RESEARCH TO IMPROVE STANDARDS OF CARE

## P160 NEUROPROTECTIVE AGENTS FOR CLINICAL TRIALS IN ALS: A SYSTEMATIC ASSESSMENT

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**Background:** Therapies that slow the progression of ALS are a crucial unmet need. Riluzole is currently the only FDA approved treatment for ALS, but its effect on survival is modest.

**Objectives:** The authors sought to identify potential neuroprotective agents for testing in phase III clinical trials and to identify which data need to be collected for each drug.

**Methods:** One hundred and twelve compounds were identified by inviting input from academic clinicians and researchers and via literature review to identify agents that have been tested in ALS animal models and in human ALS patients. The list was initially narrowed to 24 agents based on an evaluation of scientific rationale, toxicity and efficacy in previous animal and human studies. These 24 drugs then underwent more detailed pharmacological evaluation.

**Results:** Twenty drugs were finally selected as suitable for further development as treatments for ALS patients. Talampanel and tamoxifen have completed early phase II trials and have demonstrated preliminary efficacy. Other agents (ceftriaxone, minocycline, ONO- 2506 and IGF-1 polypeptide) are already in phase III trials. Remaining agents (AEOL 10150, arimoclomol, celastrol, coenzyme Q10, copaxone, IGF-1 – viral delivery, memantine, NAALADase inhibitors, nimesulide, scriptaid, sodium phenylbutyrate, thalidomide, trehalose) require additional pre-clinical animal data, human toxicity and pharmacokinetic data concerning CNS penetration prior to proceeding to large scale human testing. Further development of riluzole analogues should be considered.

**Conclusions:** Several potential neuroprotective compounds, representing a wide range of mechanisms, are available and merit further investigation in ALS.

## P161 COMPLICATIONS IN QUANTITATIVE MUSCULAR ASSESSMENT (QMA) FOR ALS PATIENTS

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**Background:** In the course of ALS, the capacity of muscle to exert force is the most direct measurement of the natural history of the disease. For this reason, muscle strength is the outcome measure that provides the most useful information in ALS clinical trials. Our previous studies concluded that outcome measures in ALS clinical trials should include QMA because the Medical Research Council Scale is insensitive to small changes.

**Objectives:** The purpose of the study was analyse possible complications in quantitative muscular assessment (QMA) in ALS patients.

**Methods:** Observational Retrospective Study. The inclusion criterion was a diagnosis of probable or definitive ALS (El Escorial diagnostic criteria). QMA for ALS patients attending Hospital Carlos III (Madrid, Spain) over two years were evaluated. The QMA protocol included measurement of shoulder abduction-adduction (supine decubitus); elbow flexion-extension (supine decubitus); knee flexion-extension (sedestation) and grip strength. Tests were performed in both limbs using an electronic strain gauge tensiometer and a electronic Jamar dynamometer. Statistical analysis was conducted, establishing the absolute and relative proportions, with SPSS 10.0 for Windows.

**Results:** One hundred and thirty-nine tests have been analysed for a total of 51 patients during the period of study. The main complications that patients presented were: fatigue post-test (86%), episode of transitory cerebral ischaemia of haemorrhagic character (0.7%; $n=1$), transitory dyspnoea (12%). Tests had to be interrupted in 5% of cases due to impossibility to maintain the supine decubitus.

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**Conclusions:** QMA are sure tests. Only in a minimum percentage of cases are the complications serious (0.7%).The most frequent complications are fatigue post-test (86%) and transitory dyspnoea (12%). Design of an informed consent for the QMA is necessary.

## P162 DESIGN OF AN INFORMED CONSENT DOCUMENT FOR QUANTITATIVE MUSCLE ASSESSMENT (QMA) IN PATIENTS WITH ALS

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**Background:** In the last 30 years, the clinical relationship between health professionals and patients has been rather modified. There are several factors that have contributed to this change: 1) Development of new techniques; 2) Recognition of patients' autonomy to decide about their own lives, health and their own bodies; 3) The increasing number of lawsuits, complaints and judgments about the problems that clinical information involves, particularly the informed consent in clinical practice.

Patients should be informed and make decisions together with the health professional on each health care assessment procedure that is carried out. An informed consent document is necessary for QMA since the tests require the patient's maximum effort and therefore they bear a series of risks or complications that the patient should know and accept.

**Objectives:** To design an appropriate informed consent document (ICD) for the realization of the QMA in patients with ALS.

**Methods:** In the elaboration of the ICD a series of stages were followed: 1) Initial design of the document by a group of health professionals ($n$=5); 2) Revision of the document for a pedagogue; 3) Focal group of patients ($n$=10); the document was explained and patient understanding was assessed; 4) The document was revised carrying out the necessary changes.

**Results:** The final text includes the following aspects: patient's personal data; name of the physiotherapist who informs; the name of the procedure that will be carried out, with simple explanation of the objective of the procedure, what it consists of and the form in which it will be carried out; a description of the known consequences of the intervention, whenever they are considered excellent or important.

There is also a description of the typical risks of the procedure and a description of the personalized risks: relating to the personal circumstances of the patient with reference to their previous state of health, age, profession, beliefs, values and attitudes, or other circumstance of a similar nature. Information can be included that makes reference to the probable nuisances of the procedure and its consequences. It is made clear that the physiotherapist will expand on the information at the patient's request.

The patient declares that they have received information about the procedure, as indicated in the previous sections, as well as of alternatives to the procedure (manual muscle assessment) and acknowledges satisfaction with the received information and of having obtained explanation on their outlined doubts, as well as of their right to withdraw the ICD at any time without expressing a reason.

A section is provided for the date and the physiotherapist's and patient's signatures. A section for the consent of a legal representative, in the event of the patient's inability is included, as is a section for the repeal of the consent.

**Conclusions:** The final ICD for the realization of the QMA is based on consent among professionals and it has been revised by the patients, improving its understanding.

## P163 THE PREVALENCE AND MANAGEMENT OF BURNING MOUTH SYNDROME IN MOTOR NEURON DISEASE

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**Background:** Burning mouth syndrome (BMS) has been described as burning sensations in the oral cavity, tongue or oral mucous membranes in the absence of clinical abnormalities (1). The most common areas in the mouth that are affected are the anterior two-thirds of the tongue, the lips and the anterior hard palate (2).

**Objective:** The aim of the study was to explore the association of burning mouth syndrome with motor neuron disease looking for causative factors and to determine treatment options.

**Methods:** MND patients attending the multidisciplinary clinic were asked about the existence of symptoms that may suggest burning mouth syndrome.

**Results:** Fourteen percent of patients reported symptoms consistent with BMS, significantly higher than the 1–5% previously reported for the general population. In these patients nutritional deficiencies such as vitamins B1, B2, B6 and zinc, and candidal infections were excluded. There were no clinical features or investigation findings to suggest Sjogrens syndrome, nor had any of the MND patients received radiotherapy. Of these patients with burning mouth symptoms, 27% had bulbar presentation. In total 73% developed bulbar involvement during the course of their MND. A trial of amitriptyline (10 mg)

was commenced in 40% of patients due to the severity of their presentation, with improvement in their symptoms.

**Conclusion:** BMS occurs in a significant proportion of MND patients. Symptoms of dysaesthesia and pain may reflect ectopic activity in denervated bulbar muscles. When severe, a trial of low-dose amitriptyline (10 mg) may be beneficial.

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## P164 OPEN-LABEL COMPARATIVE STUDY OF AMITRIPTYLINE AND DYSPORT EFFICACY IN CONTROL OF SIALORRHOEA IN ALS

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**Background:** Amitriptyline for sialorrhoea is one of the 'gold standards' in motor neuron disease (MND), but its high doses worsen quality of life due to side-effects, requiring a reduction in dose and additional approaches to treatment.

**Objectives:** We conducted an open-label comparative trial of amitriptyline and dysport efficacy in MND patients with sialorrhoea.

**Materials and methods:** We studied 10 patients with MND with bulbar involvement (eight females and two males, age range 46–69 years) diagnosed by El Escorial criteria (1998), electromyography and MRI. Five patients took 25–100 mg/d of amitriptyline (group 1), and five patients received subcutaneous dysport injections (group 2) in 250 U total dose within projection of parotids and one submandibular gland. Before and two weeks after treatment all patients underwent gravimetry of saliva (cotton pillows weighted on electronic scales Sartorius CP225D, Germany, precision 0.01 mg). Side-effects of amitriptyline were assessed by questioning whether patients had experienced at least two of the symptoms.

**Results:** Sialorrhoea assessed by gravimetry either after amitriptyline ($1.43 \pm 0.39$ and $0.91 \pm 0.61$ ml\5 min; $p < 0.05$), or dysport ($1.11 \pm 0.56$ and $0.82 \pm 0.42$ ml\5 min; $p < 0.05$) was significantly lower than before treatment. Three patients developed more then two amitriptyline side-effects. The dose was lowered (25 mg), then dysport was injected. Gravimetry two weeks later showed that sialorrhoea remained decreased ($0.56 \pm 0.25$ и $0.49 \pm 0.29$ ml\5 min; $p > 0.05$).

**Conclusions:** In cases of sialorrhoea in MND that require doses of amitriptyline causing side-effects, the lowering of amitriptyline dose and additional dysport therapy is indicated.

## P165 GASTROSTOMY IN MND: AN AUDIT OF PRE-OPERATIVE ASSESSMENT

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**Background:** Evidence-based guidelines for the management of motor neuron disease were published by the American Association of Neurologists in 1999 (1). Based on evidence from retrospective cohort studies, the following guidelines were issued: 1) PEG is indicated for patients with ALS who have symptomatic dysphagia and should be considered soon after symptom onset; 2) For optimal safety and efficacy, PEG should be offered and placed when the patient's forced vital capacity (FVC) is more than 50% of predicted.

**Objectives:** To examine local practice with respect to gastrostomy in MND: to determine whether FVC is being measured pre-operatively, and to look at mortality post-gastrostomy.

**Methods:** Audit of patient records, 1/2000 to 1/2005, for patients with MND admitted for gastrostomy to a large district general hospital with neurology services. Data were available for 22 subjects.

**Results:** Seventeen out of 22 patients underwent PEG tube placement, 5/22 RIG. Thirty-day mortality was 23%. FVC was recorded prior to gastrostomy in 13% of patients.

**Discussion:** Mortality rates are similar to published rates from the Scottish Register of Motor Neurone Disease (25% 30-day mortality (2)). Forced vital capacity is not being recorded in most cases. Clearly clinicians are not adhering to the AAN guidelines. This may reflect a reluctance of clinicians to adopt guidelines issued without a firm evidence base. However, it may reflect poor awareness of the guidelines and a lack of understanding of the role of spirometry both in determining prognosis in MND, and in informing management decisions surrounding PEG, RIG or NG tube feeding.

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## P166 DIAGNOSTIC DELAY IN MOTOR NEURON DISEASE: AN AUDIT

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**Background:** Motor neurone disease (MND) is a rapidly progressive and invariably fatal disease. Although early diagnosis is imperative, current evidence reports an average 11–22 month delay from symptom onset to diagnosis (1).

**Objectives:** In line with existing research, the MND service aims to diagnose 95% of patients within 12 months of symptom onset. This audit investigated the above standard by looking at the mean length of time taken for patients to receive a proposed and actual diagnosis of MND as well as the percentage of patients gaining a diagnosis within 12 months of symptom onset.

**Method:** We took the last 80 referrals to the MND service. Retrospective data were collected from hospital records.

**Results:** Only those patients diagnosed with MND at the regional centre were selected for inclusion in the audit ($n$=45), other cases were discarded; 68.2% had a diagnosis of MND proposed within 12 months of symptom onset and 60% had a diagnosis of MND confirmed within 12 months of symptom onset. This was below the set target of 95%. The mean time for confirmation of diagnosis was 13.3 months, which was well below the national average (1).

**Conclusions:** This audit found that while the majority of patients received a diagnosis of MND within 12 months, there were still a number of patients who did not receive their diagnosis within this time limit. There was often a considerable time delay between a proposed diagnosis being confirmed as definite MND. The wait for diagnostic neurophysiological studies and vague presenting symptoms were the most easily identifiable factors contributing to this diagnostic delay.

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## P167 TIMING OF PEG, RESPIRATORY CARE, AIDS AND ADAPTATIONS: HOW ARE PEOPLE WITH MND/ALS SUPPORTED BY THE REHABILITATION TEAMS?

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**Background:** In the Netherlands specialized ALS care teams are merged into a national network. A consensus definition of a specialized ALS care team containing eight criteria was achieved in an expert meeting. Patients with ALS receive rehabilitation treatment according to the guideline 'the rehabilitation treatment of people with ALS'. In this guideline only general recommendations regarding the timing of PEG, respiratory care and aids and adaptations are given leading to potential variability in interpretation. However; uniformity in the management of ALS concerning these aspects is in the interest of the patient with ALS.

**Objectives:** To investigate current practice of the specialized ALS care teams with respect to the timing of PEG, respiratory care and aids and adaptations.

**Methods:** A structured questionnaire was sent to the rehabilitation specialist being the representative of the ALS care teams.

**Results:** Thirty-nine (86%) of the care teams returned the questionnaire. The results showed that there is large variation in care regarding:

- PEG: Sixty-two percent of the teams use decline of respiratory function as a criterion to discuss PEG. Thirteen percent do not use weight loss as a criterion to discuss PEG. Forty-four percent discuss PEG at progressive dysarthria. In about 50% of the teams PEG insertion takes place during a 3-day hospitalization; in the remaining teams, in a daycare setting. The team member who informs and prepares a patient for PEG varies widely.
- Respiratory care: all teams discuss the possibility of NIPPV at some point in disease progression. When nocturnal hypoventilation is present 74% of the teams discuss the possibilities of ventilatory support. Progressive dysarthria is 29% of the teams' reason for discussing the possibilities of ventilatory support. Forty-three percent never or incidentally monitor a patient's vital capacity.
- Aids and adaptations: 48% of the teams reported that aids and adaptations are realised late or too late. Apart from patients' delay which is also a significant factor, two-thirds of the teams stated that local government is an impeding factor.

**Discussion and conclusions:** The results showed that there is no uniformity in the management of ALS patients with regard to PEG, respiratory care, and aids and adaptations. The Dutch guideline lacks details on these subjects. As a first step towards nationwide consensus regarding the timing of interventions in ALS, a national symposium was held in January 2005 to communicate and discuss the results of this study. The ultimate goal is to improve and standardize care of ALS patients in the Netherlands according to an evidence-based guideline.

## P168 DO ALS PATIENTS CARRY OUT PHYSIOTHERAPY SPECIFIC RESPIRATORY MUSCLE TRAINING CORRECTLY?

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**Background:** Impairments related to ALS are defined by strength loss, respiratory insufficiency, spasticity, loss of fine motor coordination, speech and swallowing difficulties. Physical therapy can potentially maximize muscle and respiratory function prolonging survival, independence in activities of daily living and quality of life. This study forms part of a larger study 'Clinical Practice Evidence- based Guidelines for Patients with ALS/MND'.

**Objectives:** The purpose of the study was to determine whether ALS patients carry out physiotherapy specific respiratory muscle training correctly.

**Methods:** Observational prospective study. The inclusion criteria were: diagnosis of probable or definitive ALS (El Escorial diagnostic criteria). ALS patients from Hospital Carlos III, Madrid, Spain were included in the study.

The 'Clinical Practice Evidence- based Guidelines for Patients with ALS/MND' include specific respiratory muscle training in the home. To determine the correct realization of the home exercises, two steps were carried out: in a first session, the patient learned and executed the exercises under the supervision of the physiotherapist; and in a second session (a month later) the realization of specific respiratory muscle training was determined without help of the physiotherapist.

**Results:** Twenty patients were assessed in the study. Only 20% correctly carried out all the programme of physiotherapy specific respiratory training a month after training. Seventy percent of the patients did not know that they were performing the exercises incorrectly. Thirty percent had doubts on the accomplishment.

**Conclusions:** ALS patients do not perform physiotherapy specific respiratory muscle training correctly. In a high percentage (80%), the execution is poor and in 70% of cases the patient does not know they are performing the exercises poorly. Therefore, periodic supervision by the physiotherapist is recommended to ensure patients carry out the respiratory exercises correctly and efficiently.

## P169 RESPIRATORY AND NUTRITITIONAL SUPPORT: A QUALITATIVE STUDY OF EXPERIENCES OF ALS PATIENTS

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**Background:** Patients with ALS all encounter severe problems with eating and breathing when the disease progresses. To reduce these problems, patients have the possibility to use respiratory and/or nutritional support. Some research has been done into the survival of patients using this kind of support and the effects on quality of life of these patients and their carers. These studies all used a quantitative methodology. To our knowledge no qualitative studies are available concerning the experiences of ALS patients with respiratory and nutritional support, and how they experience their body when non-invasive ventilation or a percutaneous endoscopic gastroscopy (PEG) is administered and is of life-saving importance.

**Objectives:** 1) To explore the experiences of ALS patients who use respiratory and/or nutritional support (non-invasive ventilation and/or a PEG) for at least three months and to focus on changes in body perception; 2) To improve the instructions given by the professional team to the ALS patient and their carers on respiratory and nutritional support and to give optimal care/guidance if patients choose and use the support; and 3) To stimulate a discussion about the ethical implications of the nutritional and respiratory support for the patients.

**Methods:** Nine patients (seven females, two males) with ALS who used respiratory and/or nutritional support for at least three months, were interviewed at home twice. A semi-structured in depth interview guide was used. All interviews were recorded on tape and transcripts were made. Transcribed interviews were analysed using the constant comparative method. Facts about disease onset, family and home situation etc., were assessed with a questionnaire prior to the interview. Functional status was assessed using the ALS Functional Rating Scale-Revised. All patients were treated in the ALS Centre of the Academic Medical Centre of Amsterdam (AMC).

**Results:** Preliminary results from the constant comparative analysis revealed that the choice for respiratory and nutritional support is, for ALS patients, often not a choice;

they feel obliged to choose the intervention in order to avoid regret afterwards. The influence of ALS, non-invasive technology and PEG on body perception is diverse. The body is perceived as a burden and a source of shame because of dependence on others and functional impairment. People with bulbar ALS reported feelings of shame in social life. The technology has influences on the perception of the body and intimate relations with others.

**Conclusions and discussion:** This qualitative research described the experiences/stories of patients with ALS using respiratory and/or nutritional support. The authors hope to stimulate a discussion about the value of qualitative research in the care for ALS clients.

## P170 THE ISSUES ON THE INDUCTION OF INVASIVE MECHANICAL VENTILATION IN JAPANESE ALS PATIENTS

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**Background:** In Japan, about 30% of ALS patients choose to use invasive mechanical ventilation (IMV). This figure is much higher than that of western countries. We may assume that one of the reasons is the Japanese way of thinking about death. We performed this study to investigate what proportion of healthy Japanese people would choose to live on IMV, if they developed ALS.

**Objective:** To investigate what proportion of the healthy Japanese population would choose to live on IMV if they had ALS.

**Method:** A questionnaire-based (Q1,Q2) study about the induction of IMV was performed on 89 medical students before and after clinical training (which included a visit to ALS patients living on home mechanical ventilation), and 25 neurologists at our institute.

Q1: If you were an ALS patient, would you choose to live with IMV?

Q2: Would your decision be different if you were able to withdraw from IMV at your own request?

**Results:** Among students before clinical practice 43.4% chose the induction of IMV; the figure dropped to 31.1% after clinical practice (they observed advanced ALS patients). Only 16.7% of neurologists chose IMV. If they could withdraw from IMV, 54.5% of students and 58.3% of neurologists chose induction of IMV.

**Discussion and conclusions:** Based on our results we assume that knowing more about ALS made students less inclined toward the induction of IMV. The percentage of induction of IMV in Japan is 30%. This figure is almost the same as the response of the students after clinical practice. Thus if patients know the situation after induction of IMV better, the figure might drop to the same level as neurologists. However, even the neurologists' response that 17% chose to wear IMV is a remarkably higher percentage compared with that of western countries. Thus, the Japanese population might have different ways of thinking about death.

The decision to choose IMV becomes more than 50% among both students and doctors if they can withdraw from IMV. In western countries the right of withdrawal from IMV is usually permitted, yet only a small percentage of patients with ALS chose IMV. In Japan the cost of IMV is covered by health insurance, so patients do not need to consider the economic burden so much. This may be one of the main reasons why so many more patients choose IMV in Japan. However, the majority of patients still do not chose IMV, because they may think they cannot accept the advanced stage of ALS and the totally locked-in state, and also because of social reasons such as difficulties of caregivers.

## P171 THE COST-EFFECTIVENESS OF EARLY NON-INVASIVE VENTILATION FOR ALS PATIENTS

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**Background:** Optimal timing of non-invasive positive pressure ventilation (NIV) initiation in patients with amyotrophic lateral sclerosis (ALS) is unknown, but NIV appears to benefit ALS patients who are symptomatic of pulmonary insufficiency. This has prompted research proposals of NIV initiated earlier in the ALS disease course in an attempt to further improve ALS patient quality of life, and perhaps survival. Prior to initiating clinical trials of novel treatments, the potential cost-effectiveness of the treatment should be considered. Cost-utility analyses allow for estimation of the cost-effectiveness of treatments in relation to their effects on quality of life, a very important outcome in ALS studies.

**Objectives:** We used a cost-utility analysis to determine a priori what magnitude of health-related quality of life (HRQL) improvement early NIV initiation would be necessary to be cost-effective in a future clinical trial. We reasoned that should the degree of improvement determined in this analysis seem plausible, future clinical trials testing early NIV would be warranted from an economic perspective. If, on the other hand, the analysis showed that an impractical degree of improvement would be necessary for the treatment to be cost-effective, future clinical trials of early NIV for ALS would be less worthwhile.

**Methods:** Using a Markov decision analytic model we calculated the benefit in health-state utility that NIV initiated at ALS diagnosis must achieve to be cost-effective. The primary outcome was the percent utility gained through NIV in relation to two common willingness-to-pay thresholds: $50,000 and $100,000 per quality-adjusted life year (QALY).

**Results:** Our results indicate that if NIV begun at the time of diagnosis improves ALS patient HRQL as little as 13.5%, it would be a cost-effective treatment. Tolerance of

NIV (assuming a 20% improvement in HRQL) would only need to exceed 18% in our model for treatment to remain cost-effective using a conservative willingness-to-pay threshold of $50,000 per QALY.

**Discussion and conclusions:** If early use of NIV in ALS patients is shown to improve HRQL in future studies, it is likely to be a cost-effective treatment. Clinical trials of NIV begun as early as the time of ALS diagnosis are therefore warranted from a cost-effectiveness standpoint.

## P172 CHARACTERISTICS OF HOSPITALIZATIONS IN AMYOTROPHIC LATERAL SCLEROSIS BETWEEN 2000 AND 2002 BASED ON A US NATIONWIDE SAMPLE

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**Background:** Patients with amyotrophic lateral sclerosis (ALS) had lengthy hospitalizations with high mortality rate in a US study for the year 1996 (1). We had previously evaluated a smaller dataset, the National Hospital Discharge Survey (NHDS) and found no significant trends in hospitalization demographics and outcomes for 1996–2002. Given that ALS is relatively rare, the NHDS dataset (ca. 270,000 records/year) captures relatively few ALS hospitalizations. We are now reporting findings based on the Nationwide Inpatient Sample (NIS, ca. 7,000,000 records/year).

**Objective:** To study the demographics and expenditure in hospitalizations of ALS patients between 2000 and 2002 based on the NIS.

**Methods:** We used the NIS, a large all-payer US inpatient care database containing data from about 1000 hospitals, approximating a 20% stratified sample of US hospital discharges. We included patients $\geq 18$ years of age. Cases were identified by ALS diagnosis (ICD code 335.20) and compared to hospitalized subjects without the ALS diagnosis. Continuous variables were compared by *t*-test, and categorical variables by Pearson's $\chi^2$ test.

**Results:** The number of ALS hospitalizations relative to the total US hospitalizations between 2000 and 2002 has been stable (22.8 to 23.2/100,000). Hospitalized ALS patients were significantly older than non-ALS patients throughout the survey period ($p < 0.01$). They were more likely to be men than non-ALS patients ($p < 0.01$). The racial and ethnic distribution was significantly different comparing hospitalized ALS from non-ALS patients ($p < 0.01$). In all years surveyed, ALS patients were more likely to be white and less likely to be black or Hispanic. The mean length of stay was twice as long for ALS patients (8.1–8.32 days) than for non-ALS patients (4.5–4.7 days). ALS admissions were more likely to be classified as emergency admissions ($p < 0.01$) and associated with high inpatient mortality ($p < 0.01$). These findings remained unchanged between 2000 and 2002. Mean hospital stay expenditure increased from 2000 to 2002 by 27% to $ 17,455 in non-ALS and by 44% to $ 33,683 in ALS patients.

**Discussion and conclusions:** Admissions in ALS patients continue to be predominantly emergency admissions. They are associated with above-average length of stay and high in-hospital mortality. The disproportionate increase in expenditure for ALS hospitalizations compared to non-ALS hospitalizations is not explained by changes in length of stay. In addition to the well established age and gender disparities between ALS and non-ALS patients, we found consistent racial and ethnic disparities in all years surveyed. Hospitalized ALS patients are more likely to be white and less likely to be black or Hispanic compared to non-ALS patients. Whether this disparity stems from socioeconomic, patient, or other factors remains to be determined.

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## P173 ALS PATIENTS' CARE: THE MND CLINIC OF MONTPELLIER. EXPERIENCE OF A MULTIDISCIPLINARY APPROACH

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**Background:** ALS is a disorder with a severe prognosis, inducing rapid and significant motor handicap. A rapid diagnosis, a systematic and complete approach for the management of the disease is needed and is expected by both the associations and the patients.

**Objectives:** To describe the organisation of our MND clinic. To present the data from one year (2004) in terms of clinical management and patient care.

**Methods:** The MND clinic, as 17 others in the country, was labelled by health authorities in 2003. The goals are: 1) precocious diagnosis; 2) systematic and homogeneous patient management; 3) organization of home care; 4) formation of care workers throughout the region; 5) organization of research. The staff is composed of three neurologists, two nurses, one psychologist-neuropsychologist, one occupational therapist, one social worker, one secretary, one technician (evaluation of the patients), and two clinical research assistants.

**Results:** A total of 320 patients, of which 120 were newly diagnosed, are being followed in the clinic. This represents a total of 1200 medical consultations, as the majority of the patients are monitored quarterly. The clinic is tightly

connected with the neuromuscular unit. False positive and false negative diagnoses of ALS remain the same between 2003 and 2004. The delay in diagnosis ranges between one week (bulbar case with dysphonia) to four years (respiratory onset with early tracheostomy and ventilation). Median survival is 48 months. Twenty percent of the patients have more than five years of evolution. Shorter survival was six months.

ALS evolution is regularly evaluated by spirometry, blood gases, weight, impédancemétrie, oxymetry, nutritional state, together with muscular testing and ALSFR-S. Five hundred consultations have been completed for both the occupational therapist and the psychologist (including 40 comprehensive cognitive evaluations). The specific case of more than two-thirds of the patients was considered by the social worker, followed by a social intervention in more than 50 cases. Research activity consists of the following programmes: 1) familial ALS network; 2) risk factors and hypoxia; 3) proteomics; 4) epidemiology and environmental risk factors; 5) cognition; 6) therapeutic trials and academic programmes (more than 150 patients included).

**Discussion and conclusion:** The Montpellier MND clinic is the second largest in the country. The rather young organisation has not yet permitted the development of protocols for care, information and a regional network for ALS care. These are our main goals for 2005–2006.

## P174 MULTIDISCIPLINARY ALS CARE IMPROVES QUALITY OF LIFE IN PATIENTS WITH ALS

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**Objective:** To examine the effect of multidisciplinary ALS care on the quality of life (QoL) in patients with ALS and their caregivers.

**Methods:** In a cross-sectional study 208 patients with ALS and their caregivers were interviewed. QoL was assessed using the 36-item Short Form Health Survey (SF-36) and two visual analogue scales (VAS). Criteria for multidisciplinary ALS care were: 1) an ALS team headed by a consultant in rehabilitation medicine and consisting of at least a physical therapist, occupational therapist, speech pathologist, dietician and a social worker; 2) use of the Dutch ALS consensus guidelines for ALS care; and 3) at least six incident ALS patients per year.

**Results:** Clinical characteristics and functional loss of the 133 patients receiving multidisciplinary ALS care and the 75 patients receiving general ALS care were similar. The percentage of patients with adequate aids and appliances was higher in those with multidisciplinary ALS care (93.1 vs. 81.3 %, $p=0.008$), while the number of visits to professional caregivers was similar in both groups. Patients in the multidisciplinary ALS care group had a better mental QoL on the SF-36 Mental Summary Score than those in the general care group ($p=0.01$). The difference in QoL was most pronounced in the domains of social functioning and mental health, and was independent of the presence of aids and appliances. No significant differences were found in the SF-36 Physical Summary Score, VAS, or in QoL of caregivers of patients with ALS.

**Conclusion:** High standard of care improves mental QoL in patients with ALS.

## P175 LIVING WITH ALS/MND: MAINTAINING PERSONAL INTEGRITY IN THE FACE OF ONGOING CHANGE AND ADAPTATION

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**Background:** The progressive death of motor neurons causes difficulties with mobility, communication, breathing and nutrition for people diagnosed with ALS/MND. While most research focuses on the disease progress, little is known of the illness experience.

**Objectives:** The aim of this study was to explore and describe what it is like to live with ALS/MND and identify how people with the disease negotiate with others for their choices in life. The purpose was to give people with the disease a rarely heard voice in qualitative research.

**Method:** A grounded theory approach was used to explicate the life world of people diagnosed and living with ALS/MND. Sample size was 25 people with the disease from rural and urban areas. Data sources were 33 transcribed in-depth interviews; written stories, prose, songs and books important to the people; and researcher field notes. The data were analysed using constant comparison analysis and managed with the software program N-Vivo.

**Results:** Analysis revealed a story of being diagnosed that was integral to understanding the illness experience of ALS/MND. Living with the disease involved the development of a decision-making process to cope with the ongoing changes that emanated from the disease's progression. The pattern was cyclic and repetitive. The basic social process that underpinned the decision-making while living with the disease was that of maintaining personal integrity. Living with ALS/MND was revealed as maintaining personal integrity in the face of ''ongoing change and adaptation''. Facets of personal integrity were feelings, image and control. Strategy types for maintaining personal integrity were protecting, regaining and sustaining. Contextual aspects that influenced the processes were past experiences of the health care system and

socioeconomic factors. The threats participants experienced as they endeavoured to maintain personal integrity were impaired communication, ''bad days'', and health professionals and service providers. Overall, participants said it was ''hard work'' to live with ALS/MND.

**Discussion and conclusions:** Laughing, denying, withdrawing and thinking creatively were important tactics used in maintaining personal integrity, but such strategies were mostly misunderstood by, or invisible to, the nurses and health care professionals who cared for the participants. As such this study revealed the hidden ALS/MND illness experience, which until now has been unacknowledged and not included in care management plans. Implications are that better understanding by health professionals of the hidden experiences will enable them to facilitate sensitive, supportive and innovative care of people with ALS/MND and their families.

**Acknowledgement:** This project was funded and supported by The Australian Research Council, Deakin University, Bethlehem Griffiths Research Foundation, Calvary Health Care Bethlehem Inc and the Motor Neurone Disease Association of Victoria. Susan King was a joint recipient of the Nina Buscombe Award 2002 and 2004.

## P176 PERSONAL EXPERIENCES OF LIVING WITH ALS/MND: ILLNESS NARRATIVES POSTED ON THE ''WEB''

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**Background:** Personal illness narratives provide powerful insights into how individuals adjust to and cope with illness. Such narratives can be obtained directly from patients, from traditionally published accounts and more recently from unpublished personal stories posted on the internet. This paper reports on one part of a study investigating published and unpublished personal narratives on ALS/MND. To date no published work has sought to investigate internet based writing about life with ALS/MND or to determine who is using this medium to publicise their illness narratives. Using the internet as a source for personal narratives presents the researcher with challenges regarding how to track down relevant information and make sense of the volume of potential material available.

**Objectives:** This paper describes the processes undertaken systematically to locate unpublished personal narratives written by people with ALS/MND posted on the internet and to identify the characteristics of the authors to establish who is using this medium to tell the story of their illness.

**Methods:** A systematic electronic search was carried out between January and March 2005 to identify and locate internet based narratives written by people with ALS/MND. Numerous search terms were employed to ensure that commonly recognized nomenclatures for the disease were incorporated. A pathway approach for the search allowed links between sites to be followed up until all links were checked and saturation was apparent. Following the application of inclusion/exclusion criteria 86 sites were accessed, from which personal writings were downloaded, archived and subject to content analysis.

**Results:** Substantially more males (76%) than females (24%) posted personal ALS/MND narratives on the internet. North America (USA 59%, Canada 21%) produced more narratives than Australia/New Zealand (11%) or Europe (9%). Authors came from a variety of occupational backgrounds. The largest occupational group (27%) consisted of manual or unskilled workers; the second largest occupational group (19%) comprised armed forces/emergency services; professional writers/academics, people from the film and music industries and health care professionals each produced 8% of the narratives. Fifty-eight sites included the author's age at diagnosis (range 20–67 years); a majority (54%) of authors were under 40 years of age at the time of their diagnosis. The experiences reported spanned the decades from the 1960s, but most authors (58%) received their diagnosis during the 1990s. Most narratives (58%) were restricted to a single entry in one section of a personal website; many (30%) were restricted to a single webpage; ongoing multiple journal entries were found on 15% of sites.

**Discussion:** This novel study demonstrates that researchers can access an invaluable source of personal narratives using a systematic approach and shows the extent to which a wide cross-section of people with ALS/MND have used new technology to publicise their personal stories.

## P177 BUILD: A VIRTUAL SUPPORT NETWORK FOR PEOPLE WITH MND

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**Background:** Communication difficulties, distance, social anxieties, and other commitments may make it difficult for some people with MND to attend 'real-world' support groups such as local support group meetings.

**Objectives:** To develop an online community for people with MND, their carers, and healthcare professionals to support one another, exchange advice, debate alternative therapies, and encourage public awareness about the condition.

**Methods:** Since 2001 a virtual message board forum has been established that is free to access for anyone

with an internet connection (www.build-uk.net) Since 2002 there has been a weekly real-time chat room for members to communicate in a safe environment. In 2005 the first Greek-language MND forum on the internet was established for use by our Greek-speaking visitors.

**Results:** Currently the site has 117 members, of whom 25 are 'regular contributors'. In the past two years the established regulars have become a crucial source of emotional support for people affected by MND including unaffected individuals at risk of inheriting the familial form of MND. One regular contributor said: ''At my local MND Association meetings, there aren't many people with MND that go. So BUILD gives me the chance to converse with other sufferers, not only from the UK but from all over the world. Also the relative anonymity afforded by a forum like BUILD allows us to discuss sensitive subjects which we would probably never discuss with anyone face to face. I think of BUILD as my internet home and the people here as my extended family.'' Another regular contributor said ''I am the youngest person at our (local) meetings and coming here means I can talk to people nearer my age as well.'' Storage of previous discussions provides an archival database of material for newly-diagnosed patients to learn more about their condition and become 'expert patients'.

**Conclusion:** The increasing use of the internet by all sectors of the MND patient population means that information and support will increasingly be delivered by high-tech means in the coming decade. The availability and anonymity of the internet means that patients are able to discuss sensitive subjects in a safe environment. Online systems also provide convenient methods of data collection and potentially clinical monitoring. Future developments may include secure virtual forums for health care professionals to form collaborations and discuss research.

## P178 INTRODUCING COMPUTER ACCESSIBILTY OPTIONS TO INDIVIDUALS WITH ALS

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**Background:** New technologies are giving individuals with motor disabilities alternative access to their computers. Information regarding these technologies is available from a variety of sources. Patients are not always aware of these sources.

**Objective:** To assess the patients' knowledge of computer accessibility in order to determine if there is a need to provide education in this area.

**Methods:** A questionnaire was administered to our clinic patients to assess their computer requirements and any difficulties they may find accessing their personal computers. They rated any difficulty they were having with their computer keyboard and mouse. Additionally, they rated their knowledge of the accessibility features currently available on the computer, as well as the software or hardware that can assist in their computer use. Information was collected on how the patients have gained their existing knowledge and what they think the proper venue to obtain future education would be. We recorded the ALSFR-S score and the patient's forced vital capacity score. In addition, the individuals rated their quality of life on a single item scale. The data were analyzed using the $t$-test ($p=0.05$).

**Results:** A total of 18 patients completed the questionnaire. Ten indicated the place to learn of computer accessibility technology would be from a health professional. We broke the data down by ALSFR-S score, specifically looking at hand function. Nine patients scored a 0 or 1 in the ALSFR-S category of handwriting. Of those individuals, seven indicated health professionals as the preferred source of information ($p=0.05$).

**Discussion and conclusions:** Our patients were reporting they would stop using their computers altogether as they began to have difficulty with access. Information is available on computer accessibility, but finding it appeared inconsistent. From this study we have learned that patients would like to receive computer accessibility information from their health professionals, yet most have relied primarily on finding it themselves. This is an unfulfilled need that the therapists can address with their patients. As a direct result of our initial survey, we have developed an educational program to address this need. This includes education on available equipment, as well as providing a trial of equipment at the clinic for interested individuals.

## P179 USE OF MODERN COMMUNICATION TECHNOLOGY BY PATIENTS WITH MOTOR NEURON DISEASE IN NORTHERN IRELAND

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**Background and objectives:** Modern communication technologies open up novel ways of managing motor neuron disease (MND) either by providing information about the disease and its symptoms by e-mail or on a website, or indeed by connecting the doctor to the patient using e-mail or video conferencing.

Whether this is feasible depends on how available these technologies are in patients' homes. To find this out we used the Motor Neurone Disease Register in Northern Ireland to ask patients by telephone a number of questions about how available modern communication technologies were to them.

**Methods:** The survey was carried out between March and May 2005. A questionnaire was sent to all 78 people on the Northern Ireland MND Register which aimed to find out information on the use of an extra telephone line, broadband or integrated services digital network (ISDN).

We asked how many people had fax machines or a computer at home and how many used e-mail and internet in general and whether they used them specifically to find information on MND. We asked them also how many thought that their relatives had found information on MND using the internet.

**Results:** Eighty-five percent of people returned the questionnaire. Of those who replied 10% had broadband or an extra line, 4% had a fax machine, 1% an ISDN line. Thirty-six percent possessed a computer with 27% using it for e-mail and 30% for the internet. Thirteen percent of people had used e-mail in connection with MND and 19% the internet. Forty-seven percent felt that their relatives had used the internet to find out about MND.

**Conclusions:** Use of e-mail and internet is remarkably low among patients with MND in Northern Ireland. Similarly, less than half of all patients' relatives had used the internet to find out information about the MND in their family. Northern Ireland has been one of the first places to make broadband universally available so there is no reason to think it is technologically backward. It is unlikely that the Motor Neurone Disease Association can increase the number of patients who are internet users and so it will need to continue to provide much of the information about MND in written form. Also, if telemedicine is to be used to deliver care to patients with MND at home, the necessary communication infrastructure will need to be introduced which will increase the cost.

## P180 QUEST (QUEBEC USER EVALUATION OF SATISFACTION WITH ASSISTIVE TECHNOLOGY) WITH COMMUNICATION AID USERS: A PILOT TRIAL IN THE UK

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**Background:** QUEST (Quebec User Evaluation of Satisfaction with assistive Technology) is an outcome measure that can give assistive technology users a way of expressing their level of satisfaction in a structured way. QUEST is a useful tool to assistive technology practitioners to help assess the usefulness of prescribed assistive equipment.

QUEST has been successfully validated for mobility products and daily living technologies but not for Electronic Assistive Technology (EAT). EAT includes Augmentative and Alternative Communication (AAC) (also known as Communication Aids) and Environmental Controls.

**Objectives:** The aim of the pilot study was to examine the views of experienced high-tech AAC device users ($n$=8). The pilot looked at how suitable QUEST 2.0 is for Communication Aid users and if modifications to QUEST 2.0 are needed for future use.

**Methods:** Participants were experienced AAC users between the ages of 18 and 65 years. All participants were able to give informed consent agreeing to participate. Participants were Communication Aid users with a variety of conditions; motor neuron disease ($n$=6), cerebral vascular accident ($n$=1) and cerebral palsy ($n$=1). Participants used a LightWRITER (n=7) or an adapted HP Jornada ($n$=1).

**Results:** Participants were broadly 'quite satisfied' with their Communication Aids. However, the majority had an area where they seemed to be less satisfied. Some questions prompted further comments and others appeared to be non-applicable to participants.

**Discussion and conclusions:** QUEST proved to be a quick, effective outcome measure that was easy to administer. The use of comments gave useful, if subjective information. Non-applicable responses also gave interesting feedback.

QUEST 2.0 appears to be a useful instrument for an AAC professional examining the satisfaction of their clients with a Communication Aid prescription. Further work to formally validate QUEST 2.0 for AAC and EAT in general is therefore desirable.

## P181 RESEARCH ON COMMUNICATION WITH AMYOTROPHIC LATERAL SCLEROSIS (ALS) PATIENTS IN A TOTALLY LOCKED-IN STATE: INTERVIEWS WITH CAREGIVERS

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**Background:** Depending on the severity of the disease, an artificial respirator can maintain life in ALS. However, as the disease progresses, paralysis of all voluntary muscles including those of the eye occurs, resulting in a totally locked-in state (TLS). At present, a device that can differentiate affirmative and negative responses based on EEG or cerebral blood flow is available. However, training to utilize this type of device is difficult, and only a handful of patients can communicate through such mechanisms in Japan.

**Objective:** The objective of the present study was to gather cues to assist in communication with patients with severe communication disorders by interviewing caregivers of ALS patients in TLS.

**Methods:** A semi-structured interview was conducted with six caregivers of four ALS patients in TLS, and the contents of the interview transcripts were categorized in order to extract related factors.

**Results:** Communication cues included such physical reactions as tachycardia, increased blood pressure, facial

flushing, and perspiration. Caregivers interpreted these reactions as signs of tension, anxiety, fear, and pain, and also attributed importance to eye positions and slight eye movements. Caregivers believed that patients were asleep if their eyes were in a superior gaze position. Moreover, they stated that although patients' eyes did not move in response to normal questions and interactions, they were sure that either the eyes moved very slightly over a period of time or that the lower eyelid contracted in response to important questions regarding therapy or social activity. Caregivers interpreted the volume of lacrimation, salivation, and tracheal secretion as indicators of happiness, sadness, and anxiety. Caregivers believed that they could read facial expressions to indicate peacefulness, laughter, and strong affirmation based on gaze; however, since these signs lack objectivity, caregivers did not base their care on their instinct and intuition.

**Discussion:** Due to limited verbal communication from patients, caregivers focused on non-verbal communication cues to ascertain emotions and patiently observed eye movements in an attempt to communicate.

## P182 NEW PARTNERSHIPS TO IMPROVE THE QUALITY OF LIFE OF PEOPLE WITH MOTOR NEURON DISEASE AND THEIR CARERS

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**Background:** Motor neuron disease (MND) has become established as a target population for palliative care services. The involvement of appropriately educated palliative care volunteers is a vital part of integrated care pathways and has the potential to reduce the acute sense of isolation resulting from MND. The usual practice of introducing volunteers during the later stages of MND can be problematic, particularly when communication difficulties impede the establishment of a constructive relationship. A new approach to palliative care volunteers is therefore required, to facilitate optimal access for people with MND and MND specific education for volunteers.

**Objectives:**

- To maximize satisfaction and well-being for both the client and the volunteer.
- To introduce comprehensively and appropriately educated volunteers to clients in the early stages of MND.
- To provide structured peer support and professional supervision to volunteers.
- To develop partnerships with existing palliative care volunteers

**Methods:** A MND volunteer visitor pilot programme was initiated. Focus groups and discussions with palliative care services helped to develop guidelines for volunteer recruitment, education and support. Existing MND information was adapted for a volunteer education programme and manual and ethical approval was obtained. Suitable MND volunteers were recruited, screened and educated and existing palliative care volunteers were invited to participate in the education programme. A range of questionnaires was developed for clients, carers and volunteers to establish the value of the education programme and the subsequent visits. People living with MND and their carers were interviewed to facilitate appropriate placement.

**Results:** The volunteers evaluated the education programme positively. A comprehensive education programme and manual for volunteers has been developed, with CD and facilitator's kit. MND volunteers have been placed with people with MND. Appropriate supervision has been provided for volunteers as well as regular peer support meetings. Evaluations of placements have been varied.

**Conclusion:** Volunteers are an important part of the integrated care pathway for people with MND. Volunteers can help people with MND achieve goals and therefore enhance quality of life. Appropriately educated volunteers may also provide a crucial link between the health care system and people with MND. This new approach provides the opportunity to introduce MND volunteers in the early stages and develop partnerships with existing palliative care services to facilitate the provision of MND specific education for palliative care volunteers.

## P183 SELF-DETERMINATION BY PATIENTS WITH ALS/MND IN JAPAN: A PRELIMINARY SURVEY

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**Background:** In Japan, a quarter of patients with amyotrophic lateral sclerosis (ALS)/motor neuron disease (MND) have been put on a mechanical ventilator with tracheostomy (TMV) at the advanced stage of the illness (1). Hayashi et al. (2) reported that 18% of patients with TMV had advanced to a totally locked-in state (TLS) at the end of the course. Currently, Japanese law prohibits anyone (including the attending physician) from discontinuing assisted ventilation once it has been started, even if the patient wishes to do so. Although there has been a lack of discussion on the way one dies in this situation, there has been considerable media information on the condition of ALS/MND at the advanced stage that may make the patients anxious, to the extent that they have concerns about their self-determination. To our knowledge, there is no published report concerning the autonomy or the self-determination of patients with ALS/MND in Japan or Asia.

**Objective:** To study the patients perception of his or her autonomy concerning options of management at the advanced stage of ALS/MND.

**Methods:** Semi-structured interviews were conducted at the homes of 14 patients with ALS/MND in the Mie prefecture of Japan. Issues explored during interview included: 1) the patients' perception of self-determination or autonomy; 2) the patients' thoughts about expressing such self-determination. These interviews, which also covered diagnosis, physical function and circumstance of care at home were carried out during the visiting survey from November 2004 to March 2005 by one of the authors (YN).

**Results:** Fourteen (10 males and 4 females) of the 21 patients surveyed were eligible for analysis on the basis of diagnosis, communication ability and cognitive function. Mean age was 57.9 years ($\pm$12.0). Nine patients seemed to poorly understand 'autonomy' ('jiritsu' in Japanese). Eleven patients answered that they wanted to exercise their right of self-determination, while the degree by which the self determination should be respected (on a percentage scale) was 89.1 ($\pm$17.6) ranging from 50 to 100%.

**Conclusions:** Many patients suffering from ALS/MND considered self-determination to be important, to choose the options for the medical management at the advanced stage of their diseases, while 'autonomy' was still unfamiliar to some Japanese patients.

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## P184 ALS PATIENTS RECEIVING HOSPICE CARE

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**Background:** The hospice is an integral aspect of ALS patient care during their end-of-life experience. Can it be assumed that hospices are familiar with the needs of ALS patients?

**Objectives:** We developed a questionnaire based on our experience with ALS patients at their end of life to determine how familiar hospice services are with these issues.

**Methods:** One hundred and fourteen sites were chosen from the National Hospice and Palliative Care Organization (NHPCO) website @ www.nhpco.org; one urban and one rural site from each state except the surrounding states of Utah where 4–6 sites were selected. Initially questionnaires were mailed, but when less than 10% were returned the questionnaires were resent by fax. When this resulted in only an additional 5% return rate, random calls to 10 hospice agencies resulted in four more returns. Most returns were incompletely filled out.

**Results:** Of 21 returns from 114 questionnaires sent, 30% reported no ALS patients in 2004, and only two agencies saw more then 15 ALS patients in 2004. Length of service (LOS) ranged from five to 232 days, compared to other diagnoses where the LOS ranged from one day to 547 days. Ten respondents felt their knowledge base of ALS was the same as other disorders; six felt it was lower and one felt it was higher as they had a registered nurse who saw all of their ALS patients. There were 16 responses to the question of whether there were cognitive changes in ALS patients and 11 said they did not see any cognitive changes and five responded "yes". The greatest challenge for hospice workers was communication with ALS patients, and the second greatest challenge was providing support to the caregiver. Equipment needs of ALS patients outweigh equipment needs of patients with other diagnoses, but clinical services offered to both groups are comparable. Symptoms consistent with upper motor neuron involvement are more prevalent in the ALS population in hospice but all other symptoms seem to be equally seen in both ALS and other diagnoses. Seventeen respondents were not aware of the American Academy of Neurology Practice Parameters, 'Care of the Patient with ALS'. One agency was aware of the standard but did not actively follow the recommendations.

**Discussion:** We were surprised at the low response rate, despite different attempts to obtain completed questionnaires. Based on the responses, hospices in general appear to have little experience with ALS patients. ALS versus other diagnosis is a small percent of the number of patients cared for in hospice and therefore there is no initiative to gain greater knowledge in this devastating disorder.

**Conclusion:** The ALS community needs to be more proactive in educating hospice agencies in the care of patients with ALS. Distributing the Practice Parameters through National Hospice Organizations is a good start. Providing educational pamphlets about the disorder and presenting at hospice conferences would help educate providers about the needs of ALS patients and maximally help them through their process of dying.

## P185 MOTOR NEURON DISEASE: PALLIATIVE CARE FROM THE ONSET

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**Background:** Within the acute setting the Hospital Palliative Care Team (HPCT) identified an unmet need for support and access to specialist palliative care services for MND patients and their carers.

**Objectives:** By developing and integrating our service with the existing MND service we aimed to facilitate access to local specialist palliative care services and

enhance the quality of care received by patients and their carers.

**Methods:** Following formal discussions, a Macmillan nurse attended a weekly MND assessment clinic (two sessions) receiving referrals for first follow-up and specific support needs from July 2002. A year later this service was increased due to the demand and complexity of referrals to three outpatient sessions involving two Macmillan nurses. This provides contact from the time of diagnosis. To date we now see all MND patients attending all three sessions. Throughout, inpatient assessment for support, symptom management and discharge planning has occurred, alongside defined telephone support for outpatients and carers. In addition the HPCT has a computer alert system informing us of readmissions to the acute setting facilitating a more rapid response from the MND team and HPCT.

**Results:** The table demonstrates access to the HPCT

| | Aug 2000 – Jun 2002 | Jul 002 – Mar 2004 | Apr 2004 – Apr 2005 |
|---|---|---|---|
| New diagnosis | 0 | 66 | 73 |
| Established disease | 3 | 46 | 93 |
| Inpatient contacts | 2 | 49 | 31 |
| Outpatient contacts | 5 | 181 | 306 |
| Referrals onto specialist palliative care services | 1 | 43 | 30 |

**Discussion and conclusions:** From the patient and carers' perspective the introduction of palliative care from the onset has been welcomed. Significant issues are discussed in a proactive and timely manner with the aim of empowering the patient, focusing on the patient's agenda and initiating thoughts of the challenges ahead. The primary care teams follow up these discussions, with access to further specialist palliative care services if needed, providing a continuity of service for the patient and improved communication between the settings. Interdisciplinary team working has developed, improving cross boundary working and communication across health, social and voluntary care professionals and settings. Development of support networks and education is evolving from this model of service delivery alongside openness and respect of roles.

## P186 END OF LIFE CARE DECISIONS IN ALS: A CROSS-ETHNIC PERSPECTIVE

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**Background:** Death and dying are extremely profound events that bring into focus important ethical, cultural, religious, and medical issues to all patients. ALS leads to progressive and often predictable decline in motor function, respiratory failure and death, while mental capacity and intellect generally remain intact in all patients. It is not known if the end-of-life care decisions differ in patients with ALS from different ethnic groups in a multi-ethnic society.

**Objective:** To study end of life decisions and clinical practices in an ethnically diverse population of patients with ALS.

**Methods:** We prospectively followed 66 patients (39 males and 27 females) of diverse ethnic groups with end-stage ALS who were seen at the Kessnich Family MDA-ALS Center and died between January 2002 and June 2004. Information with regard to patient advanced directives, health care surrogates, ALS care choices, hospice use, and palliative care was collected.

**Results:** The cohort of patients comprised 41 white Caucasians (W, 62%), 19 Hispanics (H, 29%) and six African Americans (A, 9%). Most patients anticipated death and died peacefully (93.9%) in this group. At the time of their death, 32% (W 36.6%, H 26.3%, B 20%) of them had feeding tubes, 70% (W 70.7%, H 78.9%, B 33.4%) were on non-invasive ventilatory support, and 7.6% (W 3%, H 15.5%, B 0%) had a tracheostomy and permanent ventilatory support. Advanced directives (with or without surrogate designees) were in place in 92.4% (W 95.1%, H 89.5%, B 83.3%) and were followed in 96.7% (W, 97.5%, H 89.5%, B 100%) of the patients. In this cohort, 55.5% (W 43.4%, H 68.4%, B 50%) died at home with palliative care and 41% (W 46.3%, H 26.3%, B 50%) died in hospice facility. Three patients (all W, 2.4%) died in nursing home and two patients (one each W and H) died at hospital.

**Conclusions:** These data suggest that palliative care at the end of life was relatively well managed in all ethnic groups of patients with ALS. The finding that relatively more Hispanic patients opted for permanent ventilatory support and preferred to die at home needs further study on a larger number of patients.