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International evaluation of current practices in cognitive assessment for motor neurone disease

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Background. Motor Neurone Disease (MND) is a rapidly progressive neurodegenerative disease, with up to 50% suffering from cognitive and/or behaviour changes. **Aims.** Evaluate current practices in the cognitive assessment of MND patients internationally. **Methods.** An online survey explored the use of cognitive assessments in MND clinics. **Findings.** 80/195 clinicians responded. The Edinburgh Cognitive and Behavioural ALS Screen (ECAS) was the most popular method for evaluating cognition and 72% agreed that patients screened for cognitive change have better clinical care. Thematic analysis of open-ended responses indicated that cognitive assessments help to: identify and validate changes in cognition and behaviour, aid understanding of the clinical impact of the disease, inform and direct clinical care, and infer patients' decision-making abilities. However, a number of factors affected the implementation and administration of cognitive assessments in clinics. **Conclusions.** Cognitive assessments have been implemented in MND clinics internationally and have a positive impact on clinical practice.

Introduction

Motor neurone disease (MND) is a progressive neurodegenerative disorder resulting in the loss of motor neurones from the brain, brain stem or spinal cord. It is an umbrella term used to describe variants of the disease – amyotrophic lateral sclerosis (ALS), progressive bulbar palsy (PBP), and progressive muscular atrophy (PMA). ALS affects both the upper and lower motor neurones and is the most common form of the disease, accounting for 65-85% of cases. The majority of patients present with spinal onset, whereby patients experience weakness and/or wastage in the limbs, fasciculation and spasticity. The remainder present with a bulbar-onset which is characterised by difficulties in speech (dysarthria) and swallowing (dysphagia), with a smaller proportion presenting with respiratory onset, characterised by difficulties in breathing (dystopia). It is a rapid and debilitating condition with no effective cure or treatment, and most people die within three years from the onset of symptoms (see Hardiman et al., 2017; Zarei et al., 2015 for reviews).

Although previously thought of as a disease affecting the cerebral and spinal motor neural system only, it is now widely accepted that up to 50% of people with MND experience changes in cognition and/or behaviour, with 15% meeting the criteria for frontotemporal dementia (FTD; Goldstein & Abrahams, 2013; Huynh et al., 2020). This is typically the behavioural variant (bvFTD), which is associated with early and severe changes in personality and behaviour, including apathy, disinhibition, loss of sympathy or empathy, and dietary changes (Rascovsky et al., 2011; Strong et al., 2017). The association between MND and FTD has led to the idea of a ‘disease spectrum’, with each representing one distinct end of the continuum (Lillo et al., 2012; Murphy, Henry, & Lomen-Hoerth, 2007). The remaining 35% show milder, more specific changes in cognition and/or behaviour, including executive functioning deficits (Abrahams et al., 2000; Pettit et al., 2013), language impairments (Ceslis, Argall, Henderson, McCombe, & Robinson, 2020), social cognitive deficits (Girardi, MacPherson, & Abrahams, 2011; van der Hulst, Bak, & Abrahams, 2015), and apathy (Radakovic & Abrahams, 2018). Although other behavioural changes characteristic of bvFTD can be present, and on occasion memory dysfunction is also found. Research has also indicated that patients with a *C9orf72* mutation are more likely to experience changes in cognition and behaviour, and show a distinctive neuropsychological profile compared to those without the mutation (Iazzolino et al., 2021).

Changes in cognition and behaviour may manifest as problems in communication, decision-making, passivity, and interactions with family and caregivers. Moreover, they have been shown to have a negative impact on psychological wellbeing, disease progression, treatment adherence, survival, and caregiver burden (Caga, Hsieh, Lillo, Dudley, & Mioshi, 2019; Huynh et al., 2020). It is therefore recommended that patients are given a timely cognitive assessment as such changes can have prognostic and

therapeutic consequences (Strong et al., 2017).

In the UK, the updated National Institute of Health and Care Excellence (NICE, 2016) guidelines on the assessment and management of MND state that the multidisciplinary team should be aware of the potential changes in cognition and behaviour. It is advised to explore, assess, manage and review such changes, especially when having discussions about the management of symptoms and end of life care. In practice, this would typically be done by the MND clinical nurse specialist. The publication of these guidelines subsequently prompted MND charities to provide more educational and professional development opportunities for healthcare professionals in order to raise their awareness of cognition and behavioural changes in MND. The assessment of these changes is therefore regarded as a fundamental component in the provision of appropriate care for people with MND in the UK.

Several screening and assessment tools have been developed to identify and characterise the changes in cognition and behaviour that are seen in MND (Gosselt, Nijboer, & Van Es, 2020; Simon & Goldstein, 2019). The Edinburgh Cognitive and Behavioural ALS Screen (ECAS; Abrahams, Newton, Niven, Foley, & Bak, 2014) is the most commonly used cognitive test in Scotland (Stavrou et al., 2020) and has been widely implemented in MND clinics across the UK (Hodgins, Mulhern, & Abrahams, 2020). It is a brief multi-domain assessment specifically designed for those with physical disabilities. It has been developed for use by all healthcare professionals including nurse specialists and takes approximately 20-minutes to administer.

A recent study by Hodgins et al. (2020) aimed to assess current practices of using the ECAS in the UK, and evaluate the impact it has on people with MND and those involved in their care. Assessing cognition using this method was found to have a

positive impact on patients, caregivers and healthcare professionals. Healthcare professionals reported that it helped to raise awareness of cognitive and behavioural changes, aided in the identification and understanding of such changes, and helped inform clinical decision-making. Overall, they found that the administration of a cognitive and behavioural screen improved the quality of routine clinical care in MND clinics across the UK.

Although cognitive and behavioural assessment appears to be relatively well established in MND care teams in the UK, there is a lack of knowledge on how these assessments are being used in MND clinics outside of the UK. The aim of the study was therefore to investigate current practices in assessing cognitive and behavioural changes in international MND clinics, and more specifically, to explore when and how assessments are undertaken, factors determining whether or not an assessment is carried out, and how the results affect clinical care and practice. This is of particular importance to British neuroscience nurses and other healthcare professionals looking to learn from practices in international clinics.

Materials and methods

This study was reviewed by the Department of Psychology, University of Edinburgh Ethics Committee and all participants provided their informed consent.

Major MND clinics outside of the UK were identified online via country-specific MND Associations, as well as centres listed within the European Network to Cure ALS (ENCALS), and World Federation of Neurology Research Group on MND/ALS (WFN-ALS). 195 directors, coordinators and/or principal investigators were invited via email to take part in a brief online survey about their current practices of using cognitive

assessments in MND (see Supplementary Materials). This included questions related to whether or not patients were: offered a cognitive assessment, assessed more than once, and referred to other services for further assessment. They were also asked about which assessment tests were used; which healthcare professional administered the assessments; who interpreted the results; how the results were used; and, if and how the results informed their clinical practice. The online survey was designed using Qualtrics software (Qualtrics, Provo, UT, 2020).

Data was collected between May and June 2019 and the results were a combination of quantitative closed responses (e.g. frequencies) and open-ended text responses, giving respondents the freedom and space to provide more detailed information. Descriptive statistics of the quantitative closed responses were carried out using SPSS. A Framework Analysis Method (Ritchie & Spencer, 1994) was used to thematically analyse the open-ended survey responses. The open-ended questions used in the thematic analysis included: why patients do not undergo a cognitive assessment; what determines whether or not an assessment is carried out; what determines whether or not a patient is assessed more than once; if and how the results of cognitive assessments inform clinical practice; and what would happen if a patient showed impairments in either cognition or behaviour.

A 5-step process was followed which included: familiarisation, identifying a thematic framework, indexing, charting, and mapping and interpretation. The first step of familiarisation involved author DG becoming immersed in the data by reading through the open-ended responses to develop an overview of the main ideas that emerge within the data. A theoretical framework was then developed in the second stage. The reoccurring ideas and concepts that were identified were grouped together into potential

themes in an effort to organise a conceptual framework. The third stage, indexing, refers to identifying sections of the data that are related to a specific theme. This stage was facilitated by the use of the Framework Matrices function on NVivo to link parts of the open-ended responses to the appropriate codes. Charting, the fourth stage, involved arranging and summarising the data in NVivo into columns (each representing a theme) and rows (each representing a participant). This method allowed the data to remain electronically linked to its original source to ensure that the context of the data does not disappear. The fifth and final theme of mapping and interpretation involved analysing the main characteristics as laid out in the previous step. The whole dataset and emergent themes was reviewed by author SA.

Results

Descriptive statistics of responses to closed-ended questions

80/195 (41%) responded to the survey from 25 countries (see Figure 1). Of those who responded, the majority were Neurologists (46.3%), however responses were also obtained from a range of other healthcare professionals, such as Neuropsychologists, Nurses, and Allied Health Professionals (see Table 1). The average number of MND patients seen by each centre per year was 159 ($SD = 127.7$), with an average of 62 ($SD = 67.1$) new patients being seen per year.

Nearly all participants (90%) reported that their patients underwent a cognitive assessment for clinical and/or research purposes. Of the 10% who reported that they did not administer cognitive assessments to MND patients, when asked if it would likely begin in the future, 25% answered yes - within next 12 months, 37.5% answered yes – but not within the next 12 months, and 37.5% answered no.

In MND clinics that assessed cognition, the most popular test used was the ECAS (Abrahams et al., 2014), with 66.2% reporting to use it. Over half of those using the ECAS were from European MND clinics (55.6%), while one fifth were from North American clinics (20%). The second most frequently used test was the ALS Cognitive Behavioural Screen (ALS-CBS; Woolley et al., 2010), with 32.4% reporting to use it. The majority of those (63.6%) were from North American MND clinics, and just under one third (27.4%) were from European clinics. The ALS-CBS is a short 5-minute assessment, which was designed to detect frontal lobe-mediated cognitive and behavioural changes in MND. It assesses a single cognitive domain – executive dysfunction, and includes a separate caregiver questionnaire comprised of questions assessing behavioural abnormalities common in FTD.

When questioned what the results of cognitive assessments were used for, participants selected the following optional responses: to inform and educate patients and caregivers about their difficulties (93.8%), to inform patient management/intervention (87.5%), refer to other services (64.1%), and for research purposes (62.5%; see Figure 3).

Cognitive assessments usually took place in the clinic by a neuropsychologist, neurologist or nurse specialist, although other HPs also undertook the test (OT and SLTs), and the results were often interpreted by a Neuropsychologist or Neurologist (see Table 2). The proportion of MND patients being assessed varied across clinics, with only 12.7% reporting to assess all patients (see Figure 4). When asked if patients were ever assessed more than once, 31.7% answered yes, 9.5% answered no, and 58.7% stated that only some were. The majority of participants (78.1%) reported that they refer patients to neuropsychology or psychiatry services.

When participants were asked how much they agreed or disagreed with the statement:

“ALS patients who are screened for cognitive change have better clinical care”, 72% agreed with it (see Figure 5).

Thematic analysis of responses to open-ended questions

Participants were given the opportunity to provide open-ended responses to the questions regarding determinants of whether or not: a cognitive assessment is carried out, a patient is assessed more than once, and a patient is referred to neuropsychology or psychiatry services. Questions also focussed on the ways in which results from cognitive assessments informed their clinical practice, and what they would do with regards to patient care if a patient had a specific cognitive impairment.

The participant’s responses from the open-ended questions revealed that assessing cognition and behaviour had an impact on healthcare professionals, patients and caregivers in several ways, which fall under the following overarching themes:

1) Identifies and validates changes in cognition and behaviour

The assessments ‘Provide valuable evidence of B side of the disease which is still neglected, and sometimes denied, by some families’ (Neurologist). Result could also highlight a possible biological basis such as the likelihood of C9orf72 mutation.

2) Aids understanding of the clinical impact of the disease

It helps ‘clinic staff understand patient issues’ (Neurologist) and informed them of the ‘anticipated cognitive status and possibly the progression rate’ (Neurologist).

3) Informs and directs clinical care

Indicates when a referral for specialist services is necessary, e.g. neuropsychological testing, or counselling for both the person with MND and their family.

‘The results of the ECAS can often determine if someone is referred on for more detailed neuropsychological testing’ (Research Manager).

They were also reported to promote discussion amongst healthcare professionals on the care needs of the person with MND.

‘If a patient is showing signs of cognitive issues, then I have a conversation with the physician and healthcare team in order to provide the correct course of treatment for the patient’ (Speech and Language Pathologist).

Healthcare professionals also stated that the results from cognitive assessments allow them to adjust care plans to the individual needs of the person with MND.

‘ECAS results help clinicians in better tailoring clinical and rehabilitation interventions’ (Neuropsychologist).

Results were also used to inform the suitability and recommendation of interventions.

‘Mechanical intervention...is not a good choice if patients have cognitive involvement. Thus, the results from our cognitive tests are of importance when deciding to offer invasive ventilation or not’ (Neurologist).

However some respondents questioned the clinical impact of cognitive assessments.

‘Aside from validation and education for family, whether there is a true clinical impact remains unclear to our team’ (Neurologist).

4) Infers patient’s decision-making abilities

Healthcare professionals reported that poor results on a cognitive assessment indicates issues surrounding decision-making. For example, if patients are cognitively impaired ‘they have greater difficulty understanding medical information and have trouble

making decisions’ (Medical Director) and ‘may not have the capacity to learn how to use a new piece of equipment such as a high tech communication aid or powered chair’ (Clinical Coordinator). Additionally, cognitive assessments were reported to assist with legal-related issues and have been ‘useful...when making decisions about informing regulatory authorities’ (Neurologist). Results suggesting cognitive impairment are also used to indicate when healthcare professionals need to involve family members in the decision-making process. On the other hand, one of the participants who reported that their clinic did not use cognitive assessments did not ‘buy the argument that it’s important for deciding capacity to make decisions for PEG tubes etc.’ (Clinic Director/Neurologist).

Factors affecting the implementation and administration of cognitive assessments

For those who reported doing cognitive assessments, they were often part of the clinical routine, with them being ‘offered to all patients’ (Neurologist), although on occasion some patients would refuse. Patients will also undergo an assessment if they are involved in research and it is part of the study protocol. However healthcare professionals reported that a variety of factors affected whether or not a cognitive assessment was carried out. This included the age of the patient, the disease stage, if there was a family history indicating a need for cognitive testing, and whether healthcare professionals or caregivers had concerns around the patient’s cognitive status. Furthermore, it was also reported that patients would only undergo a cognitive assessment if there was a ‘clinical need, i.e. a clear benefit to patient, family or to clarify management [such as] decision-making capacity’ (Neuropsychologist). A number of logistical barriers such as the ‘availability of neuropsychologists and

constraints of time' (Neurologist) were also identified which often prevented cognitive assessments from being carried out.

In the 10% who reported that their clinic did not administer cognitive assessments to MND patients, similar reasons were provided. The primary issue was due to a lack of staff, particularly those with expertise in assessing cognitive function. Moreover, it was often something that would only be dealt with if it became clinically necessary, otherwise they did not like 'piling on bad news' (Clinic Director/Neurologist).

Discussion

The aim of the study was to investigate current practices in assessing cognitive and behavioural changes in international MND clinics. Specifically, we aimed to explore when and how assessments are undertaken, factors determining whether or not an assessment is carried out, and how the results affect clinical care and practice. We found that cognitive assessments have been widely implemented in MND clinics internationally, with the results having a positive impact on patients, caregivers, and healthcare professionals.

Nearly every clinic (90%) reported that patients undergo a cognitive assessment for clinical and/or research purposes. The most popular tests used to assess cognition were the ECAS and ALS-CBS. Whilst a number of other standard cognitive tests were used, a limitation of these is that unlike the ECAS and ALS-CBS, they were not specifically designed for people with a physical disability. This is of importance as in MND, physical disability may manifest as problems with writing and/or speech which could potentially confound the results if not the test is not adapted for such difficulties. The most commonly used tests were often administered in clinic by a Neuropsychologist,

with the results usually being interpreted by a Neuropsychologist or Neurologist. However, it is important to note that all healthcare professionals within the multidisciplinary team can and do assess for cognition and behaviour. Previous research has shown that cognitive assessments, particularly the ECAS, have been widely implemented in MND clinics within the UK (Hodgins et al., 2020; Stavrou et al., 2020). We have expanded on this by showing that similar models appear to be used internationally. Whilst a variety of healthcare professionals are involved in administering cognitive assessments, this is not necessarily the case for interpreting the results, which appears to be limited to those with more specialist training.

When comparing the number of clinics who assessed all patients for cognitive change, we found that this was considerably lower for international MND clinics (12.7%) than for MND clinics in the UK (45%; Hodgins et al., 2020). This appeared to be explained by the number of factors that were identified to affect the likelihood of an assessment being carried out. This included clinical aspects of the patient (e.g. age, background, disease stage), whether there were concerns regarding the patients cognitive status, and if the results would be clinically useful. A small number of respondents also revealed uncertainty regarding the true clinical impact of cognitive assessments. Moreover, logistical barriers, such as limited time and the availability of trained assessors were reported to prevent cognitive assessments from being implemented into clinical practice. Although these staff-specific and resource-related barriers to assessment have been previously reported in studies exploring clinician's attitudes towards cognitive screening in MND within the UK (Crockford, Stockton, & Abrahams, 2017; Hodgins et al., 2020), this may to be a lesser extent. Less neuropsychological input in international MND clinics and the implementation of the NICE guideline recommendations for UK MND clinics may offer an explanation for these findings. Since cognitive impairment in

MND tends to be mild, there is the potential that impairments may be concealed by physical disability and go undetected in some people with the disease.

The majority of respondents (72%) agreed that MND patients who are screened for cognitive change have better clinical care, with the results of cognitive assessments being viewed as having a positive effect on patients, caregivers, and healthcare professionals. For example, they could help to identify and validate changes in cognition and behaviour, such as providing evidence that such changes can occur in the disease, and may even indicate a biological basis (e.g. C9orf72 mutation). They can aid in the understanding of the clinical impact of the disease, particularly with regards to predicting the rate of disease progression. They were also reported to inform clinical care, which included referring patients to specialist services, facilitating discussion with healthcare professionals, adapting care plans in line with the patient's individual needs, and informing the suitability and subsequent recommendation of interventions. These themes overlap with those described by Hodgins et al. (2020) in their study evaluating the clinical impact of the ECAS in the UK. We have thus extended upon their findings by showing that the results of cognitive assessments are having a comparable positive impact in MND clinics outside of the UK, which therefore validates the UK approach.

Additionally, results were often used to infer patient's decision-making abilities. It was proposed that patients with cognitive impairment were more likely to have issues surrounding decision-making, understanding information, and may not have capacity to learn new things, such as how to use new equipment. Moreover, the results were useful for healthcare professionals when deciding whether to involve family members in the decision-making process, and inform relevant

authorities about legal-related issues. This appears to be a prevalent theme as Scottish clinicians also reported that cognitive screening could help to determine a patient's mental capacity and ability to make decisions, such as power of attorney (Crockford et al., 2017). Whilst the results from a cognitive assessment may contribute towards an evaluation of a person's ability to make a specific decision, it is important to highlight that it is not an assessment of capacity and should not be used as the only source of information when drawing such conclusions (Wade, 2019).

We recognise that the majority of respondents were from European and North American countries, therefore the results may not be representative of MND clinics in other parts of the world. Furthermore, the majority of participants were Neurologists, who may not share the same views as other MND healthcare professionals. We had a response rate of 41%, therefore we also recognise that there may be a response bias for those who have incorporated cognitive assessments into their practice. Future research is therefore needed to better understand whether these results can be generalised to MND clinics outside of Europe and North America, and to other healthcare professionals. As our study only involved healthcare professionals, further work is also needed to explore the experiences of patients and their families regarding the impact that cognitive assessments have on them.

Cognitive assessment has been widely implemented into MND clinics internationally, and can have a variety of benefits for patients, caregivers, and healthcare professionals. This includes being able to: identify and validate the changes that are seen in cognition and behaviour, aid the understanding of the clinical impact of the disease, inform and

direct clinical care, and provide information regarding a person's decision-making abilities. We therefore highlight the importance of providing a timely cognitive assessment, which can identify and diagnose changes to aid patient management and may reduce caregiver burden. We recommend that this is an integral part of MND clinical care in line with NICE guidelines, and should serve as an example for incorporating assessment into other neurodegenerative disease clinics for people with other movement disorders.

Keywords

Amyotrophic lateral sclerosis; behaviour; cognition; ECAS; motor neurone disease; screening

Key points

- Screening for cognition and behaviour has been widely implemented in MND clinics outside of the UK, with the Edinburgh Cognitive and Behavioural ALS Screen (ECAS) being the most popular method used
- Cognitive assessments have a positive impact on people with MND, their caregivers and on healthcare professionals' clinical practice
- Cognitive assessments can: assist in the identification and validation of cognitive and behavioural changes, increase understanding of the clinical impact of the disease, as well as inform and direct clinical care
- Whilst results from a cognitive assessment may *contribute* towards evaluating a person's ability to make decisions, they are *not* an assessment of capacity and should not be used as the only source of information when drawing such conclusions

Reflective questions

1. How is cognition and behaviour currently screened for in MND and other neurodegenerative conditions in your practice, and how might it be improved?
2. In what ways could your multidisciplinary team address and best support patients who experience changes in their cognition and behaviour?
3. How might learning from MND clinics outside of the UK influence your own clinical practice?
4. What could healthcare professionals from MND clinics outside of the UK learn from your current practice?

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Tables with captions

Table 1: Professions of Respondents

	N (%)
Participant Occupation (N = 80)	
Allied Health Professional	7 (8.8)
Director/Clinical Care Coordinator	6 (7.5)
Doctor – Not Specified/Other	9 (11.3)
Neurologist	37 (46.3)
Neuropsychologist	7 (8.8)
Nurse	4 (5.0)
Other/Not Disclosed	10 (12.5)

Table 2 – Use of cognitive assessments

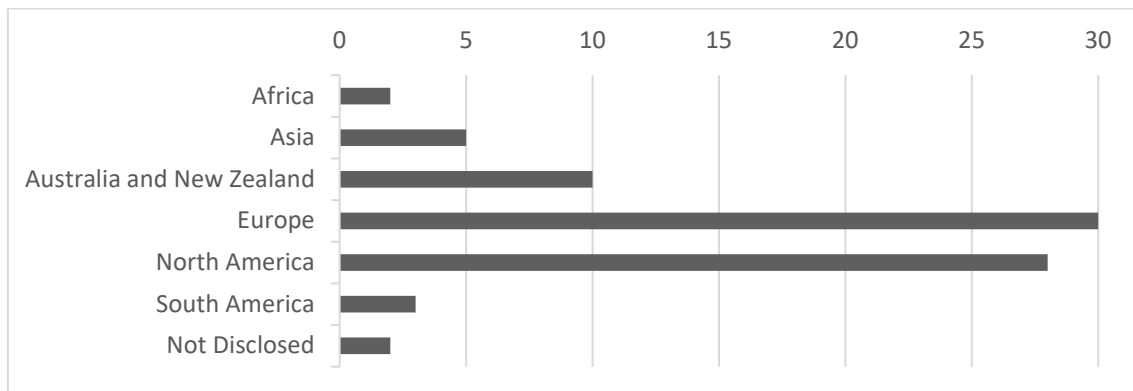
	N (%)
*Where cognitive assessments of ALS patients take place (N = 64)	
ALS Clinic	59 (92.2)
Patient's Home	12 (18.8)
Other	7 (10.9)

*Occupations who administer / interpret the results of cognitive assessments with ALS patients (N = 64)		
	Administers test	Interprets results
Clinical Neuropsychologist	34 (53.1)	36 (56.3)
Neurologist	30 (46.9)	40 (62.5)
Nurse	11 (17.2)	4 (6.3)
Occupational Therapist	6 (9.4)	6 (9.4)
Speech and Language Therapist	14 (21.9)	10 (15.6)
Other	14 (21.9)	5 (7.8)
Proportion of ALS patients that undergo a cognitive assessment in the service (N = 63)		
	All patients	New patients
5-10%	11 (17.5)	17 (27.0)
25%	15 (23.8)	7 (11.1)
50%	15 (23.8)	4 (6.3)
75%	14 (22.2)	20 (31.7)
100%	8 (12.7)	15 (23.8)

Note: Items marked * were multiple choice whereby respondents could select more than one option.

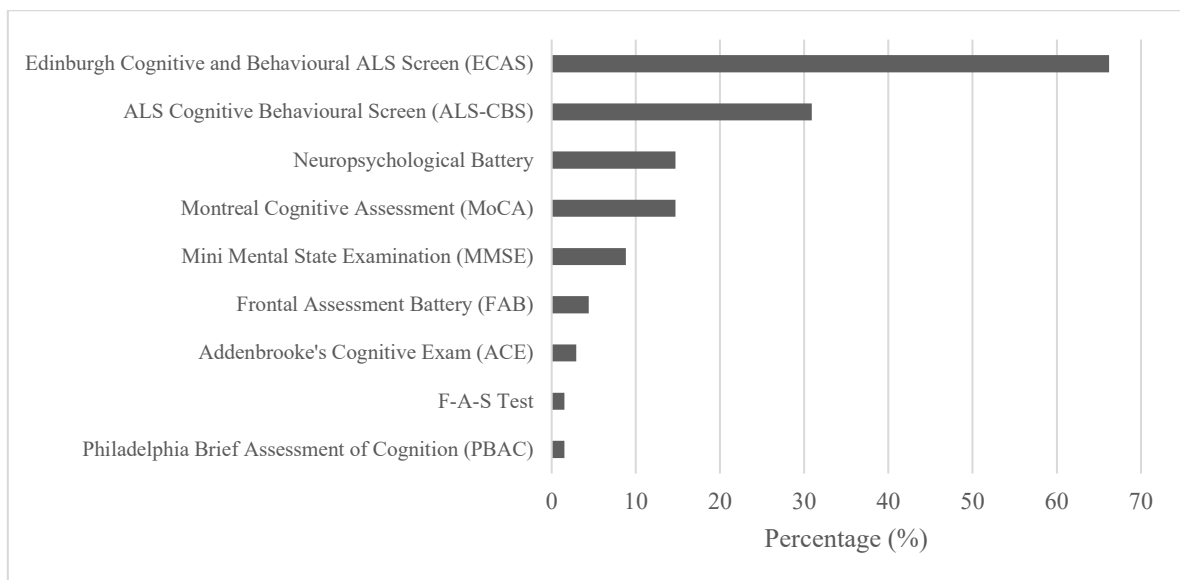
Figures with captions

Figure 1. Frequency of respondents by location



Note: N = 80 respondents. Countries included: Austria, Australia, Belgium, Bosnia and Herzegovina, Canada, China, Croatia, Colombia, Denmark, France, Germany, Greece, Ireland, Italy, Netherlands, New Zealand, Norway, Peru, Portugal, Slovenia, Spain, South Africa, Sweden, and the United States of America.

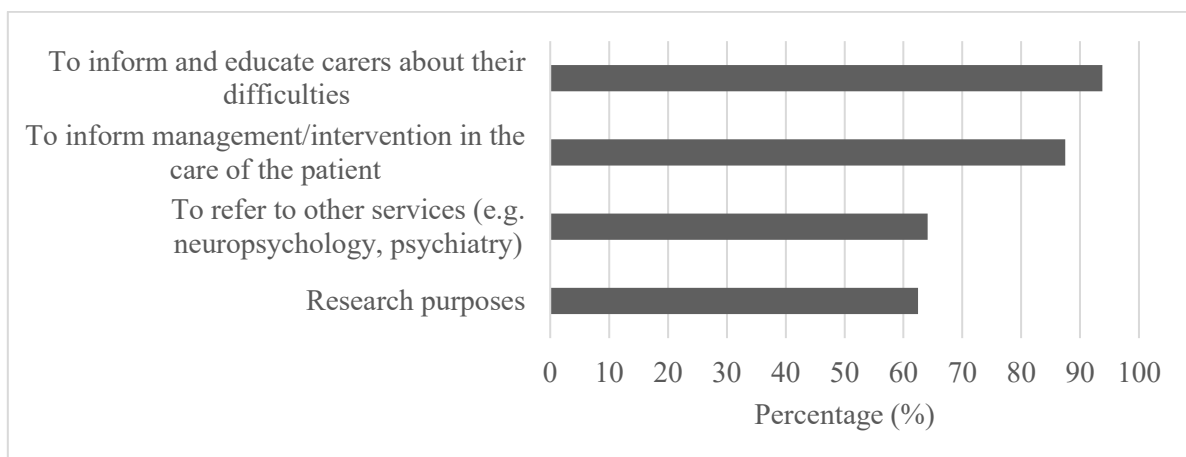
Figure 2: The method/test used to assess ALS patients for cognitive change



Note: N = 68. Participants could select multiple options and/or offer their own. **ECAS** – short multi-domain assessment design for MND (includes language, fluency, executive, memory and visuospatial). **ALS-CBS** – designed to detect frontal lobe-mediated cognitive and behavioural changes in MND and assesses a single domain (executive dysfunction). **Neuropsychological battery** – includes a range of tests to provide an overview of cognition. **MoCA** – screening instrument for detecting mild cognitive

dysfunction by assessing multiple domains (including short term memory, visuospatial abilities, executive functions, language, attention, concentration and working memory, and orientation). *MMSE* – cognitive screening test (includes tests of orientation, attention, memory, language and visuospatial skills. *FAB* – short screening test to evaluate executive function. *ACE* – used to identify cognitive impairment by assessing five cognitive domains (attention/orientation, memory, language, verbal fluency, and visuospatial skills). *F-A-S test* – assesses phonemic verbal fluency. *PBAC* – neuropsychological screening instrument which assesses five cognitive domains (working memory, visuospatial functions, language, episodic memory and comporment).

Figure 3: What cognitive assessment results were used for



Note: N = 64. Participants could select multiple options.

Figure 4: Proportion of ALS patients that undergo a cognitive assessment in the service

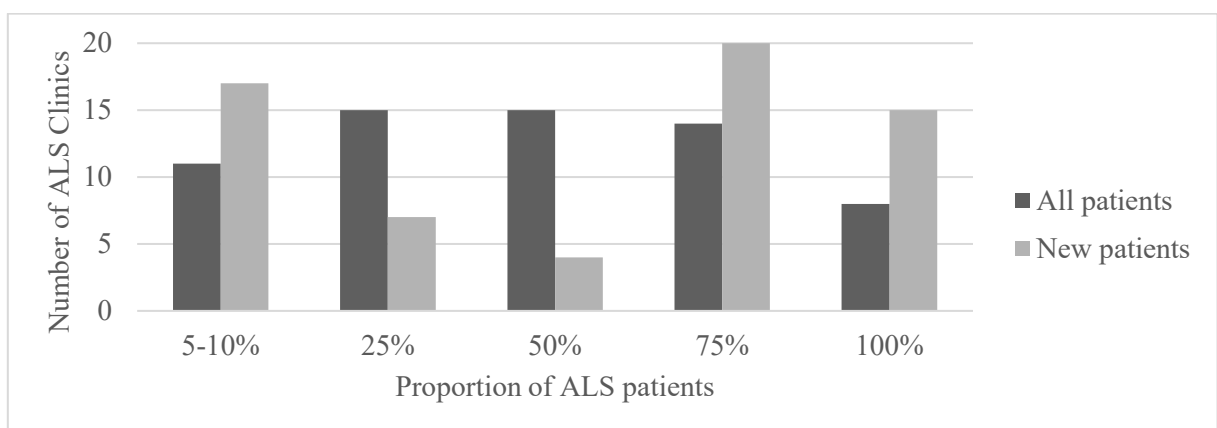
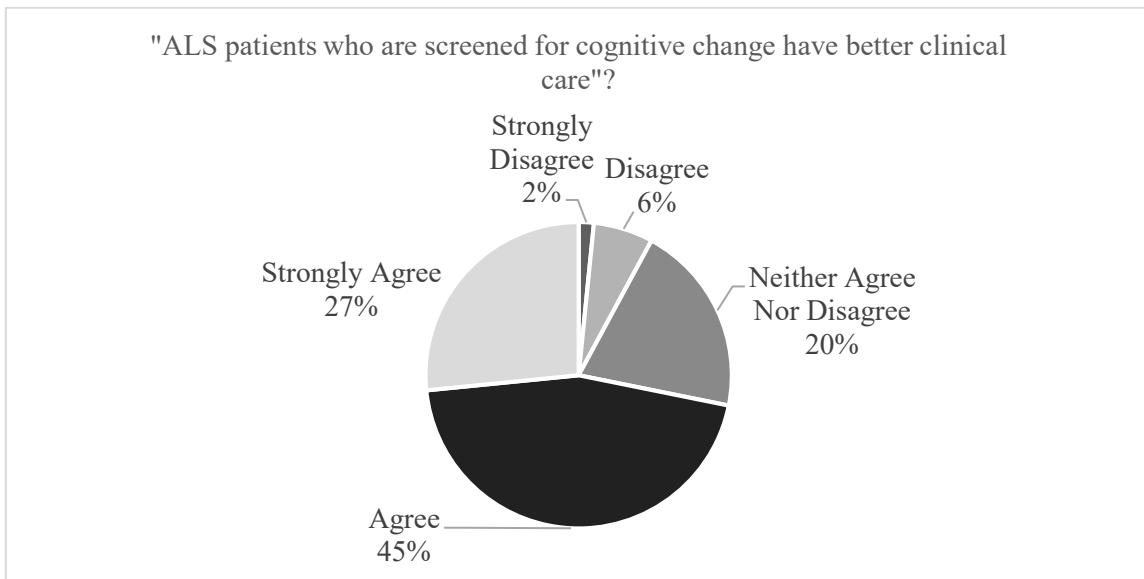


Figure 5: Impact of cognitive screening on clinical care of ALS patients



Note: N = 64