

Family carers' experiences of receiving the news of a diagnosis of Motor Neurone Disease: A national survey

Professor Samar M Aoun
Professor of Palliative Care, Associate Dean of Research
School of Nursing, Midwifery and Paramedicine
Faculty of Health Sciences
Curtin University
GPO Box U1987
Perth, WA 6845, Australia
s.aoun@curtin.edu.au

Dr Lauren J Breen
School of Psychology and Speech Pathology
Faculty of Health Sciences
Curtin University
GPO Box U1987
Perth
WA 6845
Australia
Lauren.Breen@curtin.edu.au

Dr David Oliver
Palliative Care Physician
University of Kent, Canterbury,
Kent, CT2 7NF, UK
DRDJOLIVER@GMAIL.COM

Dr Robert D Henderson
Neurologist
Royal Brisbane & Women's Hospital
Butterfield Street
Herston
Queensland 4006
Australia
Robert.Henderson@health.qld.gov.au

Dr Robert Edis
Neurologist
Sir Charles Gairdner Hospital,
Nedlands, WA 6009
Australia
robedis@it.net.au

Prof Margaret O'Connor
Professor of Nursing
Faculty of Health, Arts & Design
Swinburne University Melbourne.
Victoria 3800
Australia
margaret.oconnor@monash.edu
Ph: +61 3 9241 8441

mmoconnor@swin.edu.au

Ms Denise Howting
School of Nursing and Midwifery
Faculty of Health Sciences
Curtin University
GPO Box U1987
Perth
WA 6845
Australia
denise.howting@curtin.edu.au

Mr Rodney Harris
Chief Executive Officer
MND Victoria
PO Box 23
CANTERBURY
Victoria 3126
Australia
rharris@mnd.asn.au

Ms Carol Birks
National Executive Director
MND Australia
PO Box 430, North Sydney, NSW 2059
Australia
carolb@mndaust.asn.au

Family carers' experiences of receiving the news of a diagnosis of Motor Neurone Disease: A national survey

Abstract

Family carers have a central role in the care and support of people with MND and face the challenges of the disease from diagnosis to progression and the multiple losses of MND, but their support needs are often neglected.

This study aimed to investigate the experiences of family carers at the time of diagnosis and their satisfaction with receiving the news. An anonymous postal survey was facilitated by all MND Associations in Australia (2014) and 190 family carers completed the questionnaire. The questions centred on the SPIKES protocol for communicating bad news.

Two-thirds of family carers rated the skills of their neurologists as above average and were satisfied with the delivery of the diagnosis, in terms of having a significantly longer consultation time, the neurologist being warm and caring, satisfaction with the amount and content of information they received and relevant supports, and a plan for following up support. Conversely those who rated the neurologist's skills as below average commented on the difficulties they encountered and the long term emotional stress engendered by poor communication.

The study emphasises previous research that suggested that neurologists may require education and training in communicating the diagnosis and this should include family carers as a vital member in MND care.

Keywords: MND/ALS, family carers, breaking bad news, empathy, SPIKES protocol, MND diagnosis

1. Introduction

Motor Neurone Disease (MND), also referred to as Amyotrophic Lateral Sclerosis (ALS), is a progressive neurodegenerative disease that is always fatal [1]. There is a range of presenting symptoms, with the most common being weakness in the extremities, falls, difficulty in swallowing and speaking [2] and impairments in cognitive function and frontotemporal dementia are increasingly recognised [2, 3]. Currently, there is no effective treatment for the disease and the average time between diagnosis and death is two to three years with a small tail of long survivors [1, 4].

The disease tends to progress rapidly and family carers provide considerable support across several domains of feeding, communication, movement, and hygiene [5]. Much of the care of people with MND is provided by family carers in the home and these carers may experience a range of physical and psychological concerns such as anxiety, depression, strain, burden, fatigue, and impairments in quality of life and social contacts [6-9]. Receiving a diagnosis of MND is recognised as a central challenge for MND patients and their families [6]. In particular, issues concerning misdiagnosis [10] and dissatisfaction with the communication of the diagnosis [11-15] have been highlighted. Surveys of neurologists demonstrate that the

delivery of a diagnosis of MND is stressful and an area in which they would like more training [16, 17]. Given these issues, improving the communication of the MND diagnosis has been of increasing concern in recent years. Neurology practice guidelines underscore the challenges neurologists face in communicating MND diagnoses and provide several strategies to optimise the communication consultation, such as communicating the diagnosis in a stepwise fashion, being face-to-face in a private room without distractions; taking at least 45 to 60 minutes to convey and discuss the diagnosis; providing printed materials about MND and relevant support services to supplement the discussion; and ensuring that a follow-up appointment occurs within two to four weeks of diagnosis [18, 19].

To date there are no studies with large samples that focused separately on MND family carers' experiences of receiving the diagnosis. Typically, studies have focused on the patients' experiences of diagnosis [10, 14, 20, 21], and some combined the experiences of patients and family carers [13, 15], without due attention to the unique experiences of MND family carers. One study of caregiving experiences of current and former MND carers documented a range of support needs including respite, counselling, and access to funded and trained carers to assist them to provide care; however, they were not asked about their experiences of receiving the diagnosis [22]. Another study focused on the broader experience of support needs of a small sample (n=16) of bereaved family carers of people with MND in Australia [12]. Themes reflected the work of family carers; role changes; unremitting losses; coping mechanisms; supportive and palliative care experiences of family carers; and the experiences of receiving the diagnosis from their neurologists were poignantly mentioned: "the lack of empathy left them feeling shocked, bewildered, angry and devastated" [12, p.847]. Documenting these issues is important for two reasons. First, given that family MND carers' experiences of adverse health outcomes due to caregiving may be alleviated when their support needs are identified and addressed in a systematic and timely manner and as early as the time of diagnosis [23]. Second, the manner in which the diagnosis is communicated to families has implications for the way they adapt to the actions required for symptom management and support throughout the illness trajectory [21, 24] and through to their bereavement outcomes [12]. As such, the diagnosis of MND requires great sensitivity in the manner in which it is communicated to family carers of people with MND.

1.1 Objectives

We aimed to identify the experiences of family carers of people with MND in receiving the diagnosis, determine their overall satisfaction with the way they were given the news, and assess which aspects of the process of receiving the news were associated with greater satisfaction.

2. Methods

The methods described below are similar to those reported in the article on the patient survey [11], as the two surveys included the same questions and hence the statistical analysis was the same.

The study was approved by Curtin University Human Research Ethics Committee (HR 188/2014). The methods consisted of a cross sectional design using an anonymous postal

survey. The development of the questionnaire was undertaken after a comprehensive review of the international literature in this field and with extensive consultation with clinicians and the executive officers of the MND Associations in Australia.

2.1 Data Collection

Australian MND associations provided the number of patients on their lists who were diagnosed in the last three years and were still alive. Envelopes were mailed to each association, with each envelope containing an invitation letter bearing the letterhead of the association, one patient survey and one family carer survey with an information sheet, and a reply paid envelope. Patients and family carers were encouraged to complete the surveys independently. Carers were invited to complete the questionnaire only if they were present when the diagnosis was given to their relative/friend. MND associations attached names and address labels and posted the envelopes in their state. No further contact was made to encourage response. Data collection spanned a period from April 2014 to January 2015.

2.2 Survey instrument

The survey comprised 52 items: demographic information (age, gender, marital status, relationship to person with MND, education and postcode), date symptoms first started, date the diagnosis was first made, time spent by the neurologists giving the diagnosis. The perceived ability/skills of neurologists in delivering the diagnosis was assessed using a five-point scale from excellent to poor. Attributes of effective communication of bad news was measured by the SPIKES protocol, a well-accepted system for communicating bad news developed by Baile et al [25] and used by McCluskey et al [14]. The six domains are presented in Box 1. Each domain of the SPIKES protocol (setting, perception, invitation, knowledge, emotion and strategy) was assessed using directed questions requiring a “yes”, “no” or “do not recall” response, and directed statements requiring a response along a five point scale from “strongly agree” to “strongly disagree”. Open ended questions were included to capture more details from respondents.

Box 1: The six steps, domains and associated tasks of SPIKES

| Step | Domain | Tasks |
|-------------|---------------|---|
| 1 | Setting | Creating the right setting |
| 2 | Perception | Determining what the patient/family knows |
| 3 | Invitation | Exploring what patient/family are expecting or hoping for |
| 4 | Knowledge | Sharing the information and suggesting realistic goals |
| 5 | Emotion | Responding empathically to the feelings of patient/family |
| 6 | Strategy | Making a plan and follow through |

2.3 Analysis

The carer responses were sent back in the same reply paid envelope as the patient responses but they were coded separately. Frequencies and proportions were calculated and reported for categorical variables, and mean, standard deviation, median and range were calculated and reported for continuous and discrete variables. Normal distributions were tested using parametric means tests, and non-Normal distributions were tested using nonparametric means tests.

The SPIKES domains were analysed by calculating a summary score for each domain. There were 3 questions each in the setting and emotion domains, and 2 questions in each of the perception, emotion, knowledge, invitation, and strategy and support domains. Responses of “Yes” and “No” were coded 1 and 0, respectively. The sum of the questions in each domain was divided by the number of questions in that domain to give an average score. These scores were reported as per a continuous/discrete variable with mean, median, standard deviation and range. Responses of “do not recall” were not included in the analysis but these were few cases. The internal consistency (Cronbach’s alpha) was good to acceptable for four SPIKES domains: emotion ($\alpha=0.866$), knowledge ($\alpha=0.723$), invitation ($\alpha=0.549$), and strategy ($\alpha=0.564$).

Further analysis was also undertaken with family carers of people with MND split into two groups based upon responses to question about how they rated the ability and skills of the neurologist giving them their diagnosis: those that were rated “poor, below average or average” were assigned to one group (average or below=low rating), while those that were rated “good or excellent” were assigned to a second group (above average=high rating). Further comparisons using mean/median tests or chi-square tests were then made within these two groups according to the ability and skills rating of the neurologist to determine any differences in their experiences. Indicative responses to the open-ended questions were selected to illustrate the above and below average experiences within each domain [26].

3. Results

MND associations posted 864 questionnaire packages, with nine returned as no forwarding address was available. Responses were received from 196 family carers, of which 171 were patient-carer dyads with two carers in one instance responding for one patient, and 24 family carers with no patient responses (and 77 patients with no carer responses). Therefore, family carers in carer-patient dyads represented 88% of all responding carers. While the response rate for patients was 29% [11], it was not possible to compute a response rate for carers as we could not ascertain if all patients had family carers or if carers were not present at the diagnosis. It was a stipulated condition for the carer to be present at diagnosis in order to complete the questionnaire. Analyses were conducted on 190 family carers as six had not completed all sections of the questionnaire.

3.1 Respondents’ profile

The mean age of respondents was 62.1 years (SD=12.4, range 25-88), 67.2% were female, 93.8% were married, 82.8% were the spouses or partners of the person with MND, 11.7% were their adult children and 52.9% of family carers were retirees. The median period from diagnosis was 16 months (1-277), and median period from first symptoms to diagnosis was 11.5 months (range 1 to 240). Almost two thirds (62%) reported their care recipients having cervical/lumbar symptoms at onset, 21% had bulbar symptoms and the rest a combination of symptoms. About a third of people with MND were reported by family carers to have seen another neurologist prior to their diagnosis, 14% had seen an ENT specialist, 8% an orthopaedic surgeon, 9% a speech pathologist, and 8% a chiropractor. The majority were given the diagnosis by a neurologist (97%) through several visits: 40% had two visits, 13%

had three visits and 18% had more than 3 visits. The median length of the consultation was 40 minutes (range 10 to 200).

3.2 Ratings of neurologists' ability/skills and satisfaction with delivery of diagnosis

About two thirds of family carers (64%, n=121) rated the ability and skills of their neurologists at delivering the diagnosis as 'above average' (high rating) and 36% (n=69) rated the ability as 'average or below' (low rating). When asked to rate their satisfaction with the delivery of the diagnosis, 67% of family carers of people with MND were satisfied (very satisfied/satisfied) and 33% were not satisfied (very dissatisfied/ dissatisfied/ neither nor). Family carers' satisfaction with the delivery of diagnosis was strongly associated with the family carers' ratings of the neurologists' ability/skills [$\chi^2(1) = 88.6, p < 0.001$]. No significant differences were found in the profile between carers who were satisfied or dissatisfied.

3.3 Duration of consultation

In comparison with family carers who rated poorly the ability of their neurologists, those with high rating, had reported a similar period between first symptoms and diagnosis (mean 10 and 12 months respectively, $p=0.149$) but had significantly longer consultation times (median 45 vs 30 minutes, $p=0.002$). Family carers with high rating felt they had sufficient time taken to receive the diagnosis (just enough 85% vs 52%, and not enough 15% vs 48%, $p < 0.001$). Figure 1 demonstrates that the family carers' ratings of the neurologists' ability/skills increased as the duration of consultation increased. Similarly, Figure 2 presents the family carers' satisfaction with the delivery of diagnosis increasing as the duration of consultation increased. Both figures present comparisons with patients' findings which will be considered in the discussion section.

Figure 1: Family carers' and patients' ratings of the neurologists' ability/skills and consultation duration

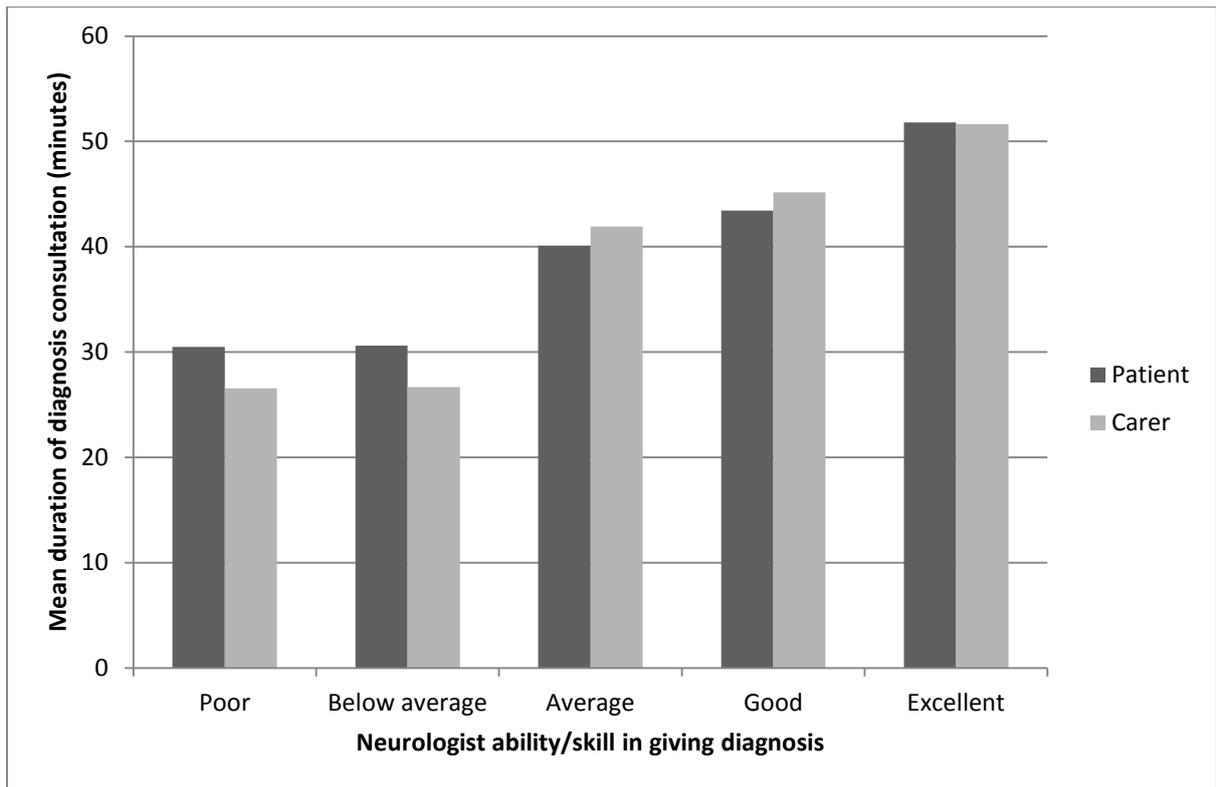
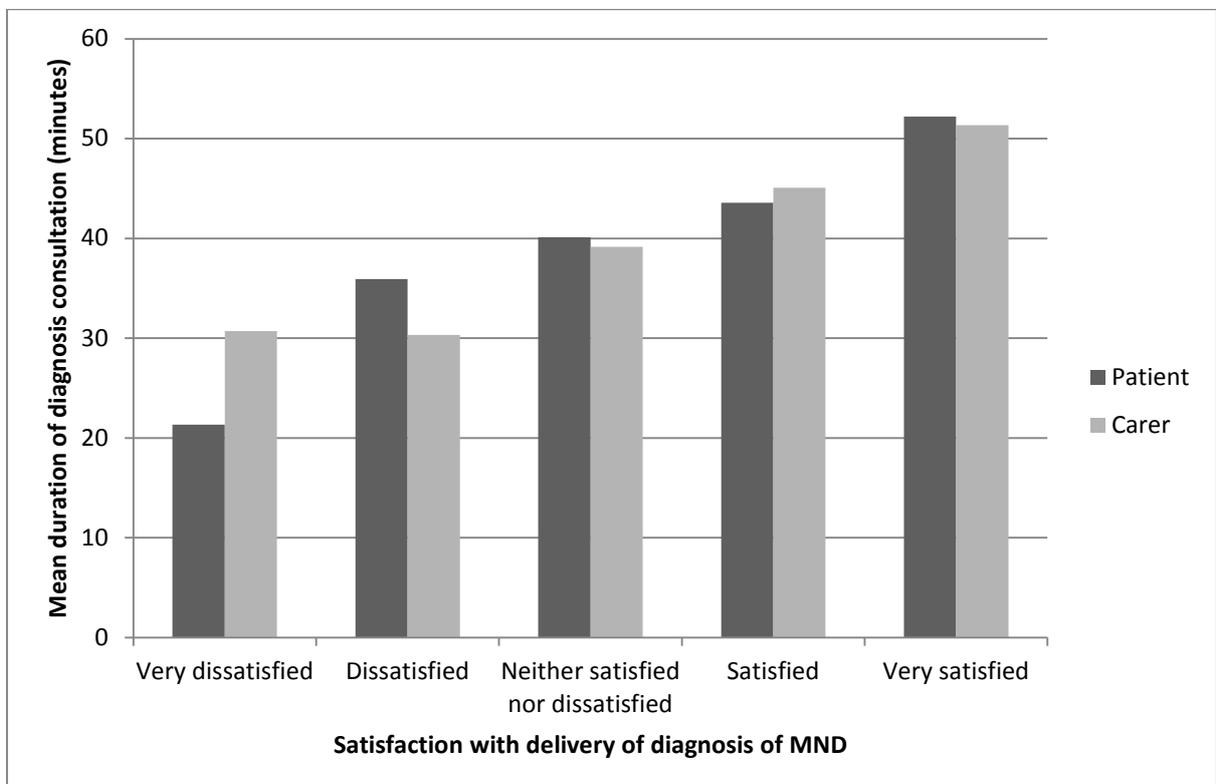


Figure 2: Family carers' and patients' ratings of satisfaction with delivery of diagnosis and consultation duration.



3.4 Comparisons within each SPIKE domain

Table 1 presents the differences in each SPIKES domain between the neurologists with high and low skill ratings in delivering the diagnosis. Where relevant, quotes are included to illustrate these differences between the two groups as experienced by the family carers.

3.4.1 Setting: Creating the right setting

The two groups of neurologists (with high and low ratings of ability) did not differ in the variables of the setting domain: the diagnosis was given in a completely private space, and there were no interruptions.

3.4.2 Perception: determining what the patient/family knows

There were no significant differences in this domain between the two groups of neurologists; in terms of the neurologist perception of the care recipient extent of knowledge of their condition, and how much detailed information they wanted to know from the neurologist.

3.4.3 Invitation: exploring what patient/family are expecting or hoping for

Family carers who rated highly the ability of their neurologists were significantly more likely to report that their care recipient was asked how much he/she knew about MND and how much detail he/she wanted to know, as seen in these positive comments:

“The neurologist explained everything in straight language and did not talk down to us respecting our knowledge.” (C115)

“I felt that my friend was totally acknowledged for her prior knowledge and strong personality with a direct, honest but not brutal assessment.” (C130)

However, the adverse effects on other family carers are evidenced in the following comment:

“As not knowing about MND, when asked what it was we were told it affects muscles, is a terminal disease and only expected to live 2 maybe 5 years at the most. This was mind blowing and numbing.” (C174)

3.4.4 Knowledge: sharing the information and suggesting realistic goals

Family carers who rated highly the ability of their neurologists were significantly more likely to be satisfied with how much detail they were provided, and were significantly more likely to be satisfied with the type of information they received. The highly rated neurologists were more likely to discuss: how the diagnosis was reached, the degree of certainty, the current state of knowledge, current research and therapeutic trials, and the Australian MND Registry. Those family carers who rated highly the ability of their neurologists were more likely to receive: further information on aspects of MND, information about MND association, and a copy of the consultation letter.

The following comments illustrate this positive experience:

"It was clear cut, not flowered up, enough info given on disease for the day as we had some knowledge, what to do next was also given." (C149)

"We were given all the relevant details we needed and were not overwhelmed with too much info until we learnt more." (C164).

"The neurologist was amazing - knew her field very well and explained all!!!." (C104)

By contrast, the negative experience was put forward as:

[Name's] neurologist at the time told him to get his affairs in order as he would probably only have 3 years to live. Very cold." (C204)

"We were literally "hit between the eyes" with the blunt way in which the diagnosis was given - but maybe this is the only way it can be handled." (C146)

"We've had great help from MND Association and from our local GPs but we've given up on the specialists who've not inspired as knowing or having researched the disease - when my wife had reaction [to] the drug supplied, it was 5 1/2 days before he returned any of our calls.." (C349)

3.4.5 Emotion: Responding empathically to the feelings of patient/family

Family carers who highly rated the ability of their neurologists were more likely to agree that their neurologist gave their loved one the diagnosis with warmth, care and empathy, that they were allowed more time to express their emotions, and they were allowed enough time to have these emotions responded to, as illustrated by these satisfied respondents:

"The neurologist was very sensitive to the enormity of the information he was giving my friend and gave her plenty of time to respond and he responded to her questions clearly, calmly and in language she could take in." (C199)

"Dr was straight forward but empathic. His suggestion that we discuss diagnosis with kids and all come back to see him with any questions in a couple of weeks. He gave us unlimited time." (C269)

By contrast, family carers who had a negative experience mentioned:

"The combination of the objective, impersonal nature of the diagnosis, shock, and ignorance of the nature of the disease, caused us to simply leave the neurologist's office very quickly. Realisation, shock, and tears set in about 10 minutes later (in a busy street)." (C227)

"1st neurologist was not very empathetic and basically said "there's nothing we can do and you're going to end up in a wheelchair" which has been a comment that doesn't disappear too easily. Always stuck. The diagnosis isn't an easy one to deliver

but perhaps a better patient manner could have been used - please express this need.” (C283)

3.4.6 Strategy: Making a plan and follow through

The following referral and support aspects were more likely to be discussed with family carers who rated highly their neurologists’ ability: a referral to the MND association, a follow-up plan for immediate and long-term support, the role of community support, the role of community palliative care, as illustrated by this satisfied respondent:

“... initial diagnosis of ALS was given and support through medication and clinic (OT, speech etc.) was explained and initiated, contact also with MND association initiated at this stage. Gene explanation and investigation began and support systems for home care etc. initiated.” (C309)

By contrast, family carers who had a negative experience reported:

“He did not refer us to MND Assoc. or offer any information on care at all. MND Association have been wonderful and advised us on care and help we can get. Without them we felt like we were alone and did not know "what's next". The regional adviser has been very helpful and a "life saver". (C133).

“I think it needs to be made clearer where and how the support will occur. It is very hard to put all the pieces together; a coordinated plan of support would be great.” (C284)

Furthermore, the neurologist was rated highly by family carers if the support was received by a neurologist or the MND specialist nurse compared to other health professionals. There was no statistical difference between the two groups in the timing of the next follow-up visit to the neurologist (about 4-7 weeks) and the median interval between subsequent follow-ups (12-16 weeks).

Table 1: Comparisons (%) within each SPIKES domain between the neurologists with high and low ratings in delivering the diagnosis

| SPIKES Domain | Survey questions corresponding to each SPIKES domains | High rating | Low rating | p-value |
|---|--|--------------------|-------------------|------------------|
| | | n=121 | n=69 | |
| Setting: Creating the right setting | Completely private space | 96.6 | 95.6 | 0.706 |
| | No interruptions | 93.2 | 85.3 | 0.120 |
| Perception: Determining what the patient/family knows | Knew some/much information about MND | 33.9 | 23.2 | 0.139 |
| | Wanted a lot/just enough detail about MND | 81.0 | 70.6 | 0.145 |
| Invitation: Exploring what patient/family are expecting or hoping for | Asked by neurologist how much they knew about MND | 54.8 | 26.1 | <0.001 |
| | Asked by neurologist how much detail they wanted to know about MND | 56.6 | 20.6 | <0.001 |
| Knowledge: Sharing | Satisfied with detail provided | 89.6 | 40.3 | <0.001 |

| | | | | |
|--|---|------|------|------------------|
| the information & suggesting realistic goals | Satisfied with type of information received | 80.7 | 19.1 | <0.001 |
| | How the diagnosis was reached | 81.0 | 62.3 | 0.006 |
| | The degree of certainty of diagnosis | 82.6 | 68.1 | 0.030 |
| | The current state of knowledge | 43.8 | 17.4 | <0.001 |
| | Current research & therapeutic trials | 29.8 | 8.7 | 0.001 |
| | The Australian MND Registry | 42.1 | 14.5 | <0.001 |
| | Receive the diagnosis in writing | 18.2 | 7.2 | 0.051 |
| | Receive further information on aspects of MND | 36.4 | 15.9 | 0.003 |
| | Information about MND Association | 56.2 | 30.4 | 0.001 |
| | MND Association publications & fact sheets | 28.9 | 17.4 | 0.083 |
| | Relevant MND internet sites | 14.9 | 13.0 | 0.831 |
| | Copy of consultation letter | 25.6 | 13.0 | 0.044 |
| | Estimate of life expectancy | 63.9 | 53.6 | 0.216 |
| Emotion: Responding empathetically to the feelings of patient/family | Diagnosis given with warmth, care & empathy | 94.2 | 26.5 | <0.001 |
| | Allowed time to express emotions | 84.0 | 20.9 | <0.001 |
| | Allowed time to have these emotions responded to by neurologist | 79.5 | 17.9 | <0.001 |
| Strategy: Making a plan & follow through | Referral to an MND multidisciplinary clinic | 41.3 | 27.5 | 0.062 |
| | Referral to the MND Association | 63.6 | 34.8 | <0.001 |
| | Role of community support | 34.7 | 7.2 | <0.001 |
| | Role of community palliative care | 23.1 | 4.3 | <0.001 |
| | Support from neurologist | 54.5 | 26.1 | <0.001 |
| | Support from MND specialist nurse | 33.9 | 14.5 | 0.004 |

3.5 Summary comparing all SPIKES domains

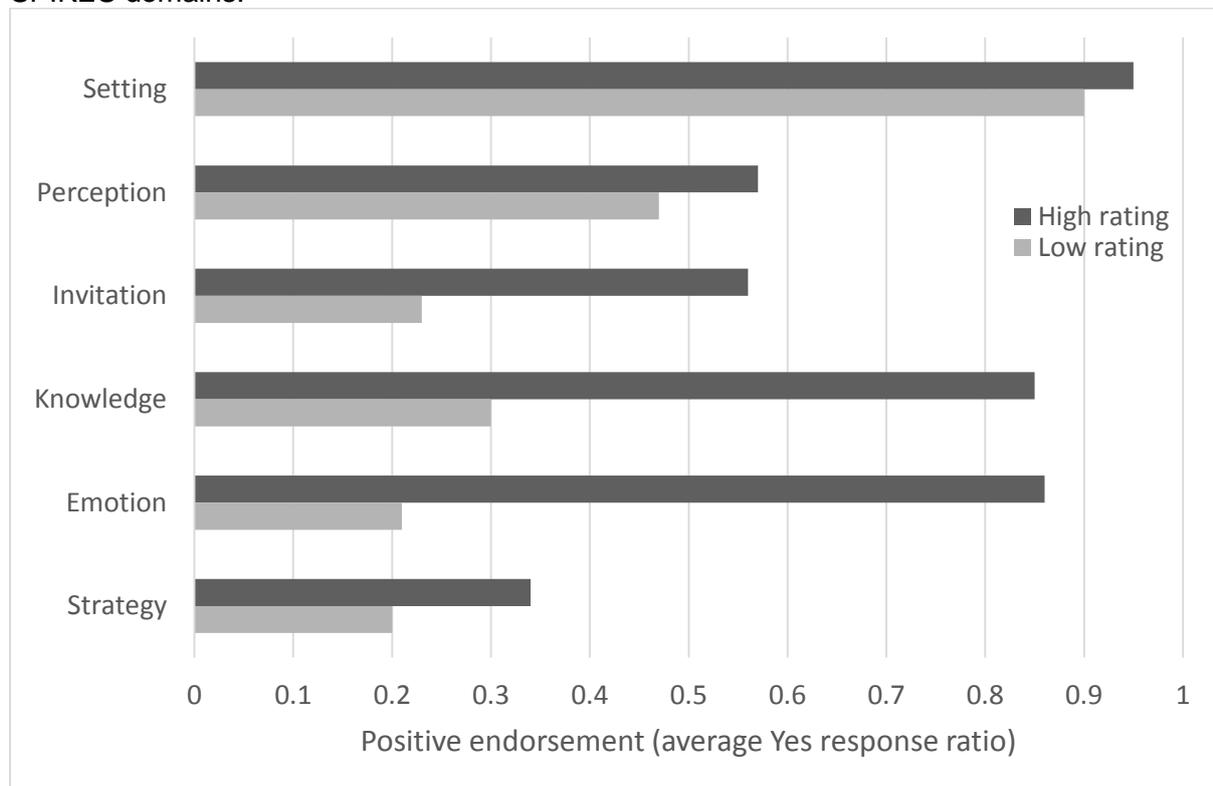
Table 2 and Figure 3 summarise the difference between the family carers' ratings of neurologists' ability (high and low ratings) across the six SPIKES domains. There were statistically significant differences in the performance ratings in delivering the diagnosis mainly across four domains, and the largest significant differences between the two groups of neurologists were in the following domains: invitation, knowledge, emotion and strategy.

Table 2: Ratings of the neurologists' ability and skills to deliver the diagnosis, grouped under the six SPIKES domains.

| | | Ability of neurologist in giving the diagnosis | | | | | |
|------------|----------------|--|---------|------------|---------|------------|----------|
| | | High rating | | Low rating | | | |
| | | N=121 | | N=69 | | Cronbach's | |
| | | | | | | p-value | α |
| Setting | Mean (SD) | 0.95 | 0.18 | 0.90 | 0.20 | 0.126 | 0.208 |
| | 95 C.I. | 0.92-0.98 | | 0.86-0.95 | | | |
| | Median (Range) | 1.0 | 0.0-1.0 | 1.0 | 0.5-1.0 | | |
| Perception | Mean (SD) | 0.57 | 0.32 | 0.47 | 0.35 | 0.056 | 0.307 |
| | 95 C.I. | 0.51-0.63 | | 0.38-0.56 | | | |

| | | | | | | | |
|------------|----------------|-----------|---------|-----------|---------|--------------|-------|
| | Median (Range) | 0.50 | 0.0-1.0 | 0.50 | 0.0-1.0 | | |
| Invitation | Mean (SD) | 0.56 | 0.40 | 0.23 | 0.35 | 0.000 | 0.549 |
| | 95 C.I. | 0.49-0.64 | | 0.14-0.31 | | | |
| | Median (Range) | 0.50 | 0.0-1.0 | 0.0 | 0.0-1.0 | | |
| Knowledge | Mean (SD) | 0.85 | 0.30 | 0.30 | 0.38 | 0.000 | 0.723 |
| | 95 C.I. | 0.79-0.90 | | 0.20-0.39 | | | |
| | Median (Range) | 1.0 | 0.0-1.0 | 0.0 | 0.0-1.0 | | |
| Emotion | Mean (SD) | 0.86 | 0.28 | 0.21 | 0.33 | 0.000 | 0.866 |
| | 95 C.I. | 0.81-0.91 | | 0.13-0.29 | | | |
| | Median (Range) | 1.0 | 0.0-1.0 | 0.0 | 0.0-1.0 | | |
| Strategy | Mean (SD) | 0.34 | 0.17 | 0.20 | 0.15 | 0.000 | 0.564 |
| | 95 C.I. | 0.31-0.37 | | 0.17-0.24 | | | |
| | Median (Range) | 0.3 | 0.0-0.8 | 0.20 | 0.0-0.7 | | |

Figure 3: Family carers' ratings of the neurologists' ability/skills grouped under the six SPIKES domains.



4. Discussion

This article has specifically focused on family carers to highlight the impact of the diagnosis on them separately from the care recipients. This distinction is important as people with MND mostly receive care at home, where their physical and psychosocial functioning is closely connected to the extent and quality of support they receive from family members who in 80% of the cases are spouses or partners, as reported in this study and the literature [27, 28]. Several previous smaller qualitative studies have described the shock of family carers with how the diagnosis was delivered [12, 15]. However this study has brought in the quantitative as well as the qualitative aspects of the experience of 190 family carers, of which a third reported that they were not satisfied with how they received the diagnosis and 36% rated the ability of the neurologist as below average. It is not surprising that these perceptions echoed those of their care recipients [11], as 88% of respondents were carer-patient dyads and only the family carers who were present at the diagnosis were invited to complete the questionnaire.

Empathy and Knowledge were the SPIKES domains where family carers had most concerns and where there were statistically significant differences between neurologists with high and low ratings of skills in communicating the diagnosis (Figure 3 and Table 1). The more empathetic neurologists gave the diagnosis with warmth and care, allowed time for the family to express their emotions and to have these emotions responded to. Family carers still expressed their feelings of shock at the lack of empathy and how the impact lingered: *“mind blowing and numbing; hit between the eyes with the blunt way; ...a comment that doesn't disappear too easily. Always stuck”*.

With the neurologists who exhibited better knowledge, family carers were satisfied with the detail and type of information provided including the information on support from the MND association and how the diagnosis was reached. The duration of the consultation was a major factor that affected satisfaction with those receiving 45 minutes being more satisfied than those who received 30 minutes, with the recommended standard of practice in international guidelines [18] being 45-60 minutes.

Notwithstanding the physical, psychological and emotional burden of the disease on MND family carers, a recent report [29] has quantified the economic disadvantage on families supporting people with MND who provide an estimate of 7.5 hours of informal care per day to people with MND: The productivity loss due to such informal care in Australia was estimated to be \$68.5 million in 2015, or \$32,728 per person with individuals shouldering most of these costs (\$44.0 million), and with government bearing the rest (\$24.5 million). Despite the many aspects of this burden, and the fact that family carers are considered co-workers in providing care and support to the care recipients [30], family carers' needs tend to be overlooked by service providers in general [31] and is reiterated in this study as depicted by the following quote:

“This diagnosis is (almost) as bad for the partner - I was hardly considered at all - little or no empathy at all. The neurologists and other doctors just don't understand

the needs of the patient's partner in order that he (or she) is better able to cope and provide support!!.” (C146)

The promotion of a collaborative approach between service providers and service users is at the heart of a person-centred approach to care with benefits to patient and carer experience and quality of care [32, 33]. The carers expressed needs for acknowledgement in this study from the time of diagnosis and throughout the caregiving journey are reinforced by previous findings, where the top priorities for support reported by MND family carers [23] included: *“Needing to know what to expect in the future; dealing with your feelings and worries and understanding your relative’s illness”*. These feelings were evident when receiving the diagnosis.

The needs of neurologists for education and training in communicating the diagnosis and responding to the emotions of patients and their families have been highlighted in a previous article [16]. It is anticipated that these programs would encompass the principles that underpin person-centred care [32], such as treating the service user with dignity, compassion, and respect, and that care is personalised, coordinated and enabling.

4.1 Limitations

The congruence in the responses between patients and carers in their overall appraisal of the neurologists’ performance provides validation of the precision of the responses between the two groups as was the case in a similar study [14]. However, there is a possibility that some carers may have provided assistance to their care recipients, thus leading to responses being similar. While we cannot ascertain a response rate for carers as some patients may not have had carers or if they did their carers may not have been present at diagnosis. Nevertheless with a patient response rate of 29% and having 88% of patient/carer dyads responding, this response rate is in line with other postal surveys which have no reminders or follow-ups sent to improve the response rate. A similar study conducted much earlier in 2004 in one American region [14] had a higher response rate of 65% and a higher dissatisfaction rate of 56%. Therefore it is likely that the dissatisfaction rate in our study would be higher than 33% if more people responded.

Other limitations include, as in the patients’ study [11], the recall bias which may not be an issue here as the median time from diagnosis was about 1.5 years, as we stipulated that only patients who were diagnosed in the last three years were invited to participate. As these questionnaires were anonymous, we cannot ascertain how many neurologists were involved with these families. However there was concordance between patients and neurologists on several key practices, such as the delivery of the diagnosis being stepwise involving two or more visits, being conveyed in a private space and the majority of patients having a family member present [16]. In Australia, the vast majority of people with MND are registered with the MND associations, therefore there may be minimal bias introduced in terms of those with bad experience through lack of follow-up not being referred to an MND association. Patients may be referred at different stages of their disease by their neurologists or other health professionals, some referred sooner than others, or they find it out themselves via the internet as per some of the comments made by the respondents.

5. Conclusions

Breaking and receiving the bad news of an MND diagnosis continues to be daunting and challenging for both neurologists and people with MND and their families. This is the first national Australian study to provide a comprehensive insight into the process of receiving the MND diagnosis from the family carers' perspective, in addition to those of the patients [11] and neurologists [16]. This study is an important contribution to the Australian landscape in terms of how well the International Guidelines on MND-ALS care [18] have been translated into clinical practice, especially the issue of how to discuss the diagnosis. These challenges are also encountered in other countries and for other terminal illnesses and therefore the findings are likely to be translatable to the international community [24, 34].

The SPIKES domains that lead to better satisfaction for patients and their family carers related to the neurologists showing more empathy and responding appropriately to their emotions, exhibiting better knowledge about detail and amount of information and available supports, providing longer consultations and referrals to MND associations. These gaps can be translated to educating neurologists and other clinicians working in this field, particularly that two-thirds of responding neurologists to a national survey expressed interest in further training in communication skills to respond to patients' emotions and development of best practice protocols [16]. This should incorporate more attention on family carers as a vital member of the MND care triad (the person, the family carer, and the health care provider) and who will be facing the challenges of ongoing care.

Acknowledgements

The authors acknowledge the financial support of The MND Research Institute Australia (MNDRIA) (Graham Lang Memorial MND Research Grant), the assistance and advice of the MND associations and the family carers of people with MND who contributed to this survey. Also special thanks for the advice of Associate Professor Paul Talman and for the assistance of Ms Kathy Deas with compiling the qualitative data.

References

- [1] J.D. Mitchell, G.D. Borasio, Amyotrophic lateral sclerosis, *Lancet* 369(9578) (2007) 2031-2041.
- [2] H. Mitsumoto, J.G. Rabkin, Palliative care for patients with amyotrophic lateral sclerosis: "prepare for the worst and hope for the best", *JAMA* 298(2) (2007) 207-216.
- [3] J. Merrilees, J. Klapper, J. Murphy, C. Lomen-Hoerth, B.L. Miller, Cognitive and behavioral challenges in caring for patients with frontotemporal dementia and amyotrophic lateral sclerosis, *Amyotrophic lateral sclerosis : official publication of the World Federation of Neurology Research Group on Motor Neuron Diseases* 11(3) (2010) 298-302.
- [4] M.C. Kiernan, S. Vucic, B.C. Cheah, M.R. Turner, A. Eisen, O. Hardiman, J.R. Burrell, M.C. Zoing, Amyotrophic lateral sclerosis, *Lancet* 377(9769) (2011) 942-955.
- [5] D. Oliver, S. Aoun, What palliative care can do for motor neurone disease patients and their families, *European Journal of Palliative Care* 20(6) (2013) 286-289.

- [6] S.M. Aoun, B. Bentley, L. Funk, C. Toye, G. Grande, K.J. Stajduhar, A 10-year literature review of family caregiving for motor neurone disease: moving from caregiver burden studies to palliative care interventions, *Palliative medicine* 27(5) (2013) 437-446.
- [7] A. Chio, A. Gauthier, A. Calvo, P. Ghiglione, R. Mutani, Caregiver burden and patients' perception of being a burden in ALS, *Neurology* 64(10) (2005) 1780-2.
- [8] L.H. Goldstein, L. Atkins, S. Landau, R. Brown, P.N. Leigh, Predictors of psychological distress in carers of people with amyotrophic lateral sclerosis: a longitudinal study, *Psychological medicine* 36(6) (2006) 865-75.
- [9] M.J. Hecht, E. Graesel, S. Tigges, T. Hillemacher, M. Winterholler, M.J. Hiltz, D. Heuss, B. Neundorfer, Burden of care in amyotrophic lateral sclerosis, *Palliative medicine* 17(4) (2003) 327-33.
- [10] J.M. Belsh, P.L. Schiffman, The amyotrophic lateral sclerosis (ALS) patient perspective on misdiagnosis and its repercussions, *J Neurol Sci* 139 Suppl (1996) 110-116.
- [11] S.M. Aoun, L.J. Breen, D. Howting, R. Edis, D. Oliver, R. Henderson, M. O'Connor, R. Harris, C. Birks, Receiving the news of a diagnosis of motor neuron disease: What does it take to make it better?, *Amyotroph Lateral Scler Frontotemporal Degener* 17(3-4) (2016) 168-78.
- [12] S.M. Aoun, S.L. Connors, L. Priddis, L.J. Breen, S. Colyer, Motor Neurone Disease family carers' experiences of caring, palliative care and bereavement: an exploratory qualitative study, *Palliative medicine* 26(6) (2012) 842-850.
- [13] L. Locock, J.B. Brown, 'All in the same boat'? Patient and carer attitudes to peer support and social comparison in Motor Neurone Disease (MND), *Soc Sci Med* 71(8) (2010) 1498-1505.
- [14] L. McCluskey, D. Casarett, A. Siderowf, Breaking the news: A survey of ALS patients and their caregivers, *Amyotrophic lateral sclerosis : official publication of the World Federation of Neurology Research Group on Motor Neuron Diseases* 5(3) (2004) 131-135.
- [15] M.R. O'Brien, B. Whitehead, B.A. Jack, J.D. Mitchell, From symptom onset to a diagnosis of amyotrophic lateral sclerosis/motor neuron disease (ALS/MND): experiences of people with ALS/MND and family carers - a qualitative study, *Amyotrophic lateral sclerosis : official publication of the World Federation of Neurology Research Group on Motor Neuron Diseases* 12(2) (2011) 97-104.
- [16] S.M. Aoun, L.J. Breen, D. Howting, D. Oliver, R. Edis, R. Henderson, M. O'Connor, R. Harris, C. Birks, Breaking the news of a diagnosis of Motor Neuron Disease: A national survey of neurologist's perspectives, *J Neurol Sci* 367 (2016) 368-374.
- [17] K.L. Schellenberg, S.J. Schofield, S. Fang, W.S.W. Johnston, Breaking bad news in amyotrophic lateral sclerosis: The need for medical education, *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration* 15(1-2) (2014) 47-54.
- [18] Anderson PM, Abrahams S, Borasio GD, de Carvalho M, Chio A, Van Damme P, et al. EFNS guidelines on the clinical management of amyotrophic lateral sclerosis (MALS)- Revised report of an EFNS task force. *Eur J Neurol.* 2012;19(3):360-375.
- [19] R.G. Miller, C.E. Jackson, E.J. Kasarskis, J.D. England, D. ForsheW, W. Johnston, S. Kalra, J.S. Katz, H. Mitsumoto, J. Rosenfeld, C. Shoesmith, M.J. Strong, S.C. Woolley, Practice parameter update: The care of the patient with amyotrophic lateral sclerosis: Multidisciplinary care, symptom management, and cognitive/behavioral impairment (an evidence-based review): Report of the Quality Standards Subcommittee of the American Academy of Neurology, *Neurology* 73(15) (2009) 1227-1233.

- [20] A. Chio, A. Montuschi, S. Cammarosano, S. De Mercanti, E. Cavallo, A. Ilardi, P. Ghiglione, R. Mutani, A. Calvo, ALS patients and caregivers communication preferences and information seeking behaviour, *Eur J Neurol* 15(1) (2008) 55-60.
- [21] H. Hugel, N. Grundy, S. Rigby, C.A. Young, How does current care practice influence the experience of a new diagnosis of motor neuron disease? A qualitative study of current guidelines-based practice, *Amyotrophic lateral sclerosis : official publication of the World Federation of Neurology Research Group on Motor Neuron Diseases* 7(3) (2006) 161-166.
- [22] M.R. O'Brien, B. Whitehead, B.A. Jack, J.D. Mitchell, The need for support services for family carers of people with motor neurone disease (MND): views of current and former family caregivers a qualitative study, *Disabil Rehabil* 34(3) (2012) 247-256.
- [23] S.M. Aoun, K. Deas, L.J. Kristjanson, D.W. Kissane, Identifying and addressing the support needs of family caregivers of people with motor neurone disease using the Carer Support Needs Assessment Tool, *Palliative & supportive care* (2016) 1-12.
- [24] S. Connolly, M. Galvin, O. Hardiman, End-of-life management in patients with amyotrophic lateral sclerosis, *Lancet Neurol* 14(4) (2015) 435-442.
- [25] W.F. Baile, R. Buckman, R. Lenzi, G. Glober, E.A. Beale, A.P. Kudelka, SPIKES—A Six-Step Protocol for Delivering Bad News: Application to the Patient with Cancer, *Oncologist* 5(4) (2000) 302-311.
- [26] K. Malterud, Qualitative research: standards, challenges, and guidelines, *Lancet* 358(9280) (2001) 483-488.
- [27] G. Bruletti, L. Comini, S. Scalvini, R. Morini, A. Luisa, M. Paneroni, M. Vitacca, A two-year longitudinal study on strain and needs in caregivers of advanced ALS patients, *Amyotroph Lateral Scler Frontotemporal Degener* 16(3-4) (2015) 187-95.
- [28] F. Pagnini, P. Banfi, C. Lunetta, G. Rossi, G. Castelnuovo, A. Marconi, F. Fossati, M. Corbo, E. Molinari, Respiratory function of people with amyotrophic lateral sclerosis and caregiver distress level: a correlational study, *BioPsychoSocial medicine* 6(1) (2012) 14.
- [29] Deloitte Access Economics, The economic value of informal care in Australia in 2015: Carers Australia., Deloitte Access Economics, Barton, ACT, 2015.
- [30] G. Ewing, L. Austin, J. Diffin, G. Grande, Developing a person-centred approach to carer assessment and support, *British journal of community nursing* 20(12) (2015) 580-4.
- [31] S. Aoun, K. Deas, C. Toye, G. Ewing, G. Grande, K. Stajduhar, Supporting family caregivers to identify their own needs in end-of-life care: Qualitative findings from a stepped wedge cluster trial, *Palliative medicine* 29(6) (2015) 508-17.
- [32] A. Collins, Measuring what really matters. Towards a coherent measurement system to support person-centred care., The Health Foundation, London, 2014.
- [33] Health Foundation, Person-centred care made simple. What everyone should know about person-centred care., The Health Foundation, London, 2014.
- [34] Oliver D, Borasio G, Caraceni A et al. A consensus review on the development of palliative care for patients with chronic and progressive neurological disease. *European Journal of Neurology* 23 (2016) 30-38.