

ARTICLE

Receiving the news of a diagnosis of motor neuron disease: What does it take to make it better?

SAMAR M AOUN¹, LAUREN J BREEN², DENISE HOWTING¹, ROBERT EDIS³,
DAVID OLIVER⁴, ROBERT HENDERSON⁵, MARGARET O'CONNOR⁶,
RODNEY HARRIS⁷ & CAROL BIRKS⁸

¹*School of Nursing, Midwifery and Paramedicine*, ²*School of Psychology and Speech Pathology Faculty of Health Sciences, Curtin University, Perth, Western Australia*, ³*Royal Perth Hospital, Shenton Park Campus, Shenton Park, Australia*, ⁴*Wisdom Hospice and University of Kent, Rochester, UK*, ⁵*Royal Brisbane & Women's Hospital, Herston, Queensland*, ⁶*Faculty of Health, Arts & Design, Swinburne University Melbourne, Victoria*, ⁷*MND Victoria, Canterbury, Victoria*, and ⁸*MND Australia, Gladesville, Australia*

Abstract

Our objectives were to identify the experiences of people with MND in receiving the diagnosis and to determine which aspects of breaking this bad news were associated with greater satisfaction with the way the diagnosis was delivered to them. An anonymous postal survey was facilitated by all MND associations in Australia, in 2014, and centred on the SPIKES protocol for communicating bad news. Of the patients ($n=248$, response rate 29%), 36% were dissatisfied with the delivery of the diagnosis and gave low ratings on the ability/skills of their neurologists to deliver the diagnosis. It was evident that the longer the patients spent with their neurologists during breaking such bad news, the more they were satisfied and the higher they rated the neurologists' abilities/skills. The largest significant differences between neurologists rated as having high or low skills in delivering the diagnosis were in four domains: 1) responding empathically to the feelings of patient/family; 2) sharing the information and suggesting realistic goals; 3) exploring what patient/family are expecting or hoping for; and 4) making a plan and following through. In conclusion, with over one-third of patients dissatisfied with their experience, there is room for improvement in the practice of neurologists in specified areas that could form the basis for changing practice, and the development of standards and protocols likely to have implications at the international level.

Key words: *MND/ALS, breaking bad news, empathy, SPIKES protocol, MND diagnosis*

Introduction

Motor neuron disease (MND), also known as amyotrophic lateral sclerosis (ALS) or Lou Gehrig's disease, is a progressive neurodegenerative disease. Causes are unknown for approximately 90% of people with MND, and all cases are fatal with no effective treatment affecting the underlying disease mechanism (1). Presenting symptoms vary but typically include weakness in the hands or feet, trips and falls, swallowing difficulties, and slurred speech and nearly half of all people with MND may have impaired cognitive function, although overt frontotemporal dementia is less commonly observed (2,3). The time between diagnosis and death averages two to three years and most people with MND die from respiratory failure (1,4). In population

studies, approximately 10–20% of patients have a prolonged survival (5). The psychosocial impact of MND is intensified by the rapid speed of deterioration and relentless losses experienced by people with MND and their families (6–8).

Receiving a diagnosis of MND is challenging for patients and their families and is akin to an existential shock (7,9). The manner in which patients learn of a serious diagnosis is central to good practice guidelines in healthcare and underpins protocols developed for communicating bad news (10,11). Practice guidelines for neurologists acknowledge the challenges they face in communicating diagnoses of MND and emphasise communicating the diagnosis face-to-face in a private room without distractions; providing adequate time, at least 45–60 min, for conveying the diagnosis and

its discussion; complementing the face-to-face discussion with printed materials about the disease and relevant support services; and establishing a follow-up appointment that occurs within two to four weeks of diagnosis (12,13). Thus, the diagnosis is envisaged to be made in a way that acknowledges the individual's emotional and spiritual needs as well as addressing the medical and practical needs (14).

The communication of an MND diagnosis has implications for the way that patients and families move from the news of their diagnosis to the actions required for support throughout the illness trajectory; however, few studies have investigated communication of the diagnosis from the perspectives of people receiving the news (15). A survey of people attending a specialist MND centre in Italy demonstrated that most respondents were satisfied with the communication of the diagnosis (16). However, other studies demonstrate less satisfactory experiences. A survey of 64 people with MND in the United States showed that 27% reported at least one misdiagnosis before the MND diagnosis, with 8% undergoing costly and unnecessary surgeries (17). A more recent American survey of 144 people with MND demonstrated that fewer than half were satisfied with the way the diagnosis was communicated and 16% characterized it as poor (18). These issues with communication of the diagnosis feature prominently in qualitative studies exploring experiences of people with MND and their family caregivers. Interviews with 24 people with MND, 18 family caregivers, and 10 bereaved caregivers concluded that the delivery of the diagnoses ranged from being communicated in an informative and sensitive manner to being communicated abruptly, without empathy, and in public spaces (19). In an Australian study involving interviews with 16 bereaved family caregivers of people with MND, the caregivers described an absence of compassion during delivery of the diagnosis, which had long-standing effects (6). To date there are no studies with large sample sizes that have been conducted to ascertain the nature and extent of these challenging experiences.

We aimed to identify the experiences 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.

Methods

The study was approved by Curtin University Human Research Ethics Committee. 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.

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 containing an invitation letter bearing the letterhead of the association, a patient survey with an information sheet, and a reply paid envelope. 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.

Survey instrument

The patient survey consisted of 51 questions: demographic information (age, gender, marital status, 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 were assessed using a 5-point scale from excellent to poor. Attributes of effective communication of bad news were measured by the SPIKES protocol, a well-accepted system for communicating bad news developed by Baile et al. (10) and used by McCluskey et al. (18). The six domains are: 1) Setting – establishing the appropriate setting; 2) Perception – determining the needs and the perception of the patient; 3) Invitation – requesting an invitation to give the news; 4) Knowledge – providing knowledge and information to the patient; 5) Emotion – exploring the patient's feelings; 6) Strategy – forming a strategy with the patient to go forward. Each domain of the SPIKES protocol (setting, perception, invitation, knowledge, emotion and strategy) was assessed using direct questions requiring a 'yes', 'no' or 'do not recall' response, and directed statements requiring a response along a 5-point scale from 'strongly agree' to 'strongly disagree'. Open ended questions were included to capture more details from respondents.

Analysis

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 non-parametric means tests.

The SPIKES domains were analysed by calculating a summary score for each domain. There were three questions in each of the setting and emotion domains, and two 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 particularly good for three SPIKES domains: emotion ($\alpha = 0.897$), knowledge ($\alpha = 0.731$) and invitation ($\alpha = 0.636$).

Further analysis was also undertaken with 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 χ^2 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 (20).

Results

MND associations posted 864 questionnaires, with nine returned as no forwarding address was available. Responses were received from 248 patients, yielding an overall response rate of 29% (ranging from 35% to 26% between five Australian states). Comparative analyses were conducted on 243 patients as five had missed completing a few sections of the questionnaire.

Respondents' profile

The mean age of respondents was 66.4 years (SD = 11.0, range 30–91 years), 59% were male, 78% were married, and 75% were retirees. The median period from diagnosis was 15 months (1–87), period from first symptoms to diagnosis was 10 months for the median (range 1–119). Over two-thirds (69%) reported having cervical/lumbar symptoms at onset, 19% had bulbar symptoms and the rest a combination of symptoms. About one-third of people with MND had seen another neurologist prior to their diagnosis, 15% had seen an ENT specialist, 11% an orthopaedic surgeon, 9% a speech pathologist, and 8% a chiropractor. The majority were given the diagnosis by a neurologist (95%) through several visits: 33% had two visits, 17% had three visits and 19% had more than three visits. The median length of the consultation was 30

min (range 1–300 min). Seventy percent of patients reported that they had a relative present with them during the consultation.

Ratings of neurologists' abilities/skills and satisfaction with delivery of diagnosis

About two-thirds of patients (64%, $n = 156$) rated the abilities and skills of their neurologists at delivering the diagnosis as 'above average' (high rating) and 36% ($n = 87$) rated the ability as 'average or below' (low rating). When asked to rate their satisfaction with the delivery of the diagnosis, 65% of people with MND were satisfied (very satisfied/satisfied) and 35% were not satisfied (very dissatisfied/dissatisfied). Patients' satisfaction with the delivery of diagnosis was strongly associated with the patients' ratings of the neurologists' abilities/skills ($\chi^2(1) = 88.7, p < 0.001$).

Duration of consultation

Patients who rated highly the abilities of their neurologists had a shorter period between first symptoms and diagnosis (mean 20 and 26 months, respectively, $p = 0.021$), had significantly longer consultation times (median 40 vs. 30 min, $p < 0.001$), felt they had sufficient time taken to receive diagnosis (just enough 84% vs. 48%, and not enough 4% vs. 34%, $p < 0.001$), and were males (64% vs. 49%, $p = 0.042$). Figure 1 demonstrates that the patients' ratings of the neurologists' abilities/skills increased as the duration of consultation increased. Similarly, Figure 2 presents the patients' satisfaction with the delivery of diagnosis increasing as the duration of consultation increased.

Comparisons within each SPIKE domain

Table I presents the differences in each SPIKES domain between the neurologists with high and low ratings in delivering the diagnosis.

Setting: creating the right setting. The two groups of neurologists (with high and low ratings of ability) significantly differed in two out of the three variables of the setting domain; the diagnosis was given in a completely private space and there were no interruptions.

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's perception of the patient's extent of knowledge of their condition and how much detailed information they wanted to have from the neurologist.

Invitation: exploring what patient/family are expecting or hoping for. Patients who rated highly the abilities of their neurologists were significantly more likely to be asked how much they knew about MND and how much detail they wanted to know.

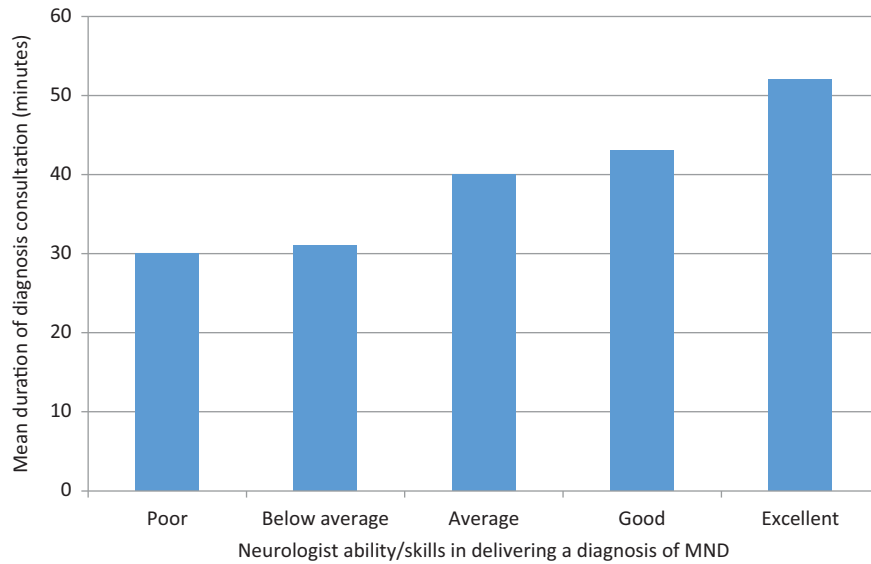


Figure 1. People with MND ratings of the neurologists' abilities/skills and consultation duration.

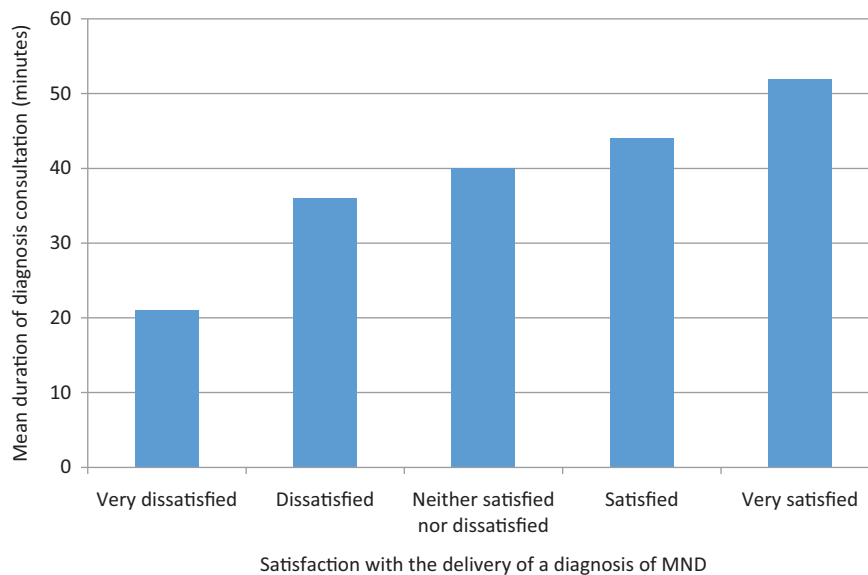


Figure 2. People with MND ratings of satisfaction with delivery of diagnosis and consultation duration.

Knowledge: sharing the information and suggesting realistic goals. Patients who rated highly the abilities 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 patients who rated highly the abilities of their neurologists were more likely to receive the diagnosis in writing, further information on aspects of MND, information about MND associations, MND association publications and fact sheets, relevant MND internet sites, a copy of

the consultation letter and more likely to receive an estimate of their life expectancy.

Emotion: responding empathically to the feelings of patient/family. Patients who highly rated the abilities of their neurologists were more likely to agree that their neurologist gave them 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.

Strategy: making a plan and follow-through. The following referral and support aspects were more likely to be discussed with patients who rated highly their neurologists' abilities: a referral to an MND multidisciplinary clinic, a referral to the MND association, a follow-up plan for immediate and

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

SPIKES domain		High rating <i>n</i> = 156	Low rating <i>n</i> = 87	<i>p</i> -value
Setting: creating the right setting	Completely private space	98	91	0.019
	No interruptions	95	86	0.027
	Relative/friend present	72	70	0.653
	Seen alone	19	24	0.412
Perception: determining what the patient/ family knows	Knew some/much information about MND	34	28	0.389
	Wanted a lot/just enough detail about MND	86	79	0.147
Invitation: exploring what patient/family are expecting or hoping for	Asked by neurologist how much they knew about MND	54	24	<0.001
	Asked by neurologist how much detail they wanted to know about MND	40	13	<0.001
Knowledge: sharing the information and suggesting realistic goals	Satisfied with detail provided	82	45	<0.001
	Satisfied with type of information received	76	31	<0.001
	How the diagnosis was reached	71	56	0.024
	The degree of certainty of diagnosis	76	61	0.013
	The current state of knowledge	42	22	0.002
	Current research and therapeutic trials	21	7	0.003
	The Australian MND Registry	39	17	<0.001
	Receive the diagnosis in writing	21	8	0.011
	Receive further information on aspects of MND	31	14	0.003
	Information about MND Association	54	21	<0.001
	MND Association publications and fact sheets	32	12	<0.001
	Relevant MND internet sites	18	3	0.001
	Copy of consultation letter	24	13	0.031
Estimate of life expectancy	64	48	0.020	
Emotion: responding empathetically to the feelings of patient/family	Diagnosis given with warmth, care and empathy	88	29	<0.001
	Allowed time to express emotions	84	28	<0.001
	Allowed time to have these emotions responded to by neurologist	83	26	<0.001
Strategy: making a plan and follow- through	Referral to an MND multidisciplinary clinic	44	22	<0.001
	Referral to the MND Association	51	23	<0.001
	Role of community support	29	8	<0.001
	Role of community palliative care	21	8	0.011
	Support from neurologist	50	28	0.001
	Support from MND specialist nurse	32	16	0.007

long-term support, the role of community support and the role of community palliative care. Furthermore, the neurologist was rated highly by patients 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 seven weeks) and the median interval between subsequent follow-ups (12 weeks).

Summary comparing all SPIKES domains

Table II and Figure 3 summarize the difference between the patients' ratings of neurologists' abilities (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 III presents reported experiences of people with MND within each SPIKE domain, with respondents' quotes depicting positive and negative experiences categorized by ratings of neurologists' skills in delivering the diagnosis.

Discussion and conclusion

This is the first national Australian study to provide a comprehensive insight into the process of receiving an MND diagnosis from the patients' perspective. Previous studies in Australia and elsewhere were qualitative, relied on small samples and portrayed mainly disaffected patients due to the self-selection bias (6,19,21).

Table II. Ratings of the neurologists' abilities and skills to deliver the diagnosis, grouped under the six SPIKES domains.

SPIKES domains	Rating of abilities of neurologists by people with MND				<i>p</i> -value	Cronbach's α
	High rating		Low rating			
	<i>n</i> = 156		<i>n</i> = 87			
Setting					0.031	0.470
Mean \pm SD	0.90	± 0.16	0.83	± 0.22		
Median (Min, Max)	1.0	(0.3, 1)	1.0	(0.3, 1)		
Perception					0.127	0.290
Mean \pm SD	0.60	± 0.29	0.54	± 0.30		
Median (Min, Max)	0.5	(0, 1)	0.5	(0, 1)		
Invitation					<0.001	0.636
Mean \pm SD	0.47	± 0.43	0.19	± 0.31		
Median (Min, Max)	0.5	(0, 1)	0.0	(0, 1)		
Knowledge					<0.001	0.731
Mean \pm SD	0.79	± 0.34	0.38	± 0.43		
Median (Min, Max)	1.0	(0, 1)	0.0	(0, 1)		
Emotion					<0.001	0.897
Mean \pm SD	0.85	± 0.31	0.26	± 0.38		
Median (Min, Max)	1.0	(0, 1)	0.0	(0, 1)		
Strategy					<0.001	0.473
Mean \pm SD	0.32	± 0.17	0.20	± 0.15		
Median (Min, Max)	0.3	(0, 0.9)	0.1	(0, 0.7)		

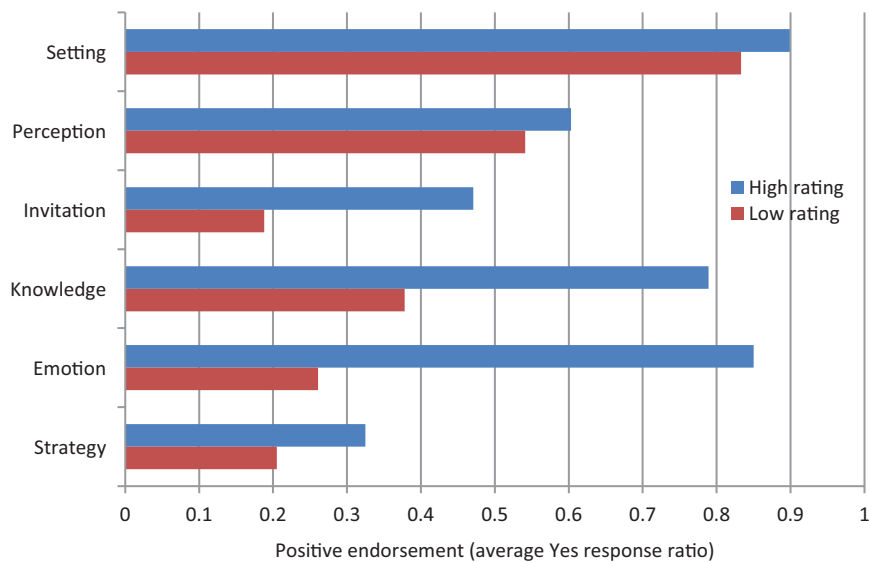


Figure 3. People with MND ratings of the neurologists' abilities/skills grouped under the six SPIKES domains.

Respondents to this national survey seemed to fall into two distinct groups: 64% were satisfied with the delivery of the diagnosis and rated highly the abilities/skills of their neurologists, and 36% were dissatisfied with the delivery of the diagnosis and gave low ratings on the abilities/skills of their neurologists. The qualitative comments of people with MND in Table III significantly reinforce the positive and negative experiences, which were closely related to the perceived ability of their neurologist and the reported satisfaction with the diagnosis delivery process. These comments are not dissimilar to those obtained from the smaller qualitative studies and particularly the comments about the need for empathy (6,19,21).

Additionally, the overall rating of skills was closely associated with performance within the SPIKES domains. In particular, the largest significant differences in ability/skills in delivering the diagnosis between the two groups of neurologists according to the patients were in four domains: 1) Emotion (responding empathically to the feelings of patient/family), where empathy was an important attribute of highly rated neurologists and how they dealt with the emotions of the patient and family; 2) Knowledge (sharing the information and suggesting realistic goals) where highly rated neurologists gave information about all aspects related to the disease, certainty, research, estimation of life expectancy and information on the MND association; 3) Invitation

Table III. Respondents' feedback depicting their experiences with the two groups of neurologists in each SPIKE domain.

SPIKE domains	Rating	Quotes illustrating positive and negative experiences categorized by ratings of neurologists' skills and satisfaction with delivery of diagnosis (high and low ratings).
SETTING (Creating the right setting) A completely private space	Low	"Info given to me only referred to negatives which upset me - at time of info given no private area to remain in and went out to public area that was full and I was upset in front of them." (P190)
Presence of interruptions	Low	"I was not impressed by the number of interruptions during my neurologist consultations i.e. Phone calls, missing reports, and copies of reports being brought in..." (P340)
Presence of a relative or friend at the diagnosis	High	"Neurologist was very professional but also empathetic and answer(ed) question from both patient and carer plus two adult children" (P225)
	Low	"I should have been told to bring my husband or a support person when he knew he was giving me the diagnosis." (P230)
PERCEPTION (determining what the patient/family knows) Perception for just enough knowledge needed	High	"At initial diagnosis I did not want to know long-term details. About three months later I wanted to know long-term details." (P370)
	Low	"Explanation too fast and too technical despite my reasonable biological knowledge." (P201)
INVITATION (exploring what patient/family are expecting or hoping for) Prior knowledge about MND	High	"I had been told by my first neurologist that MND was on the table so I had done some research in the months between him and seeing neurologist... It was all pretty grim so I did not delve too deeply". (P09)
	Low	"I think they should start with 'What do you know about MND?' As an icebreaker." (P229)
Details wanted	High	"The Dr. was very kind, sympathetic. He asked how much I wanted to know." (P73)
	Low	"At the first interview I should have been given the opportunity to be asked if I would like more information or not. The internet is a daunting place and that is where I had to source information." (P205)
KNOWLEDGE (sharing the information and suggesting realistic goals) Satisfaction with how much detail was provided	High	"I was not bombarded with the whole story in one hit and allowed time to deal with it." (P296)
	Low	"I was told very bluntly about my situation and how it would basically progress. He said if I had any plans do them now." (P176)
Satisfaction with type of information received	High	"Information given also by way of DVD - excellent!" (P94) "The neurologist spoke clearly, calmly and answered my questions in 'layman's' language." (P162)
	Low	"Would gladly have driven into a pole on the way home because I was given no hope." (P64)
Discussion of how diagnosis was reached	High	"Not enough information given, certainly at the first two appointments. His presentation could best be described as robotic." (P205)
	High	"... I asked whether it could be MND and she replied that that was what she was thinking. She then explained the reasons for her diagnosis to my wife and me, and explained the associated implications very clearly and with empathy." (P152)
The degree of certainty	Low	"Ist visit to neurologist said he would like to do nerve conduction tests. He didn't say why. I asked could I have an ageing disease like MS. He was surprised that I asked and said 'No, I suspect motor neuron disease.'" (P234)
	High	"Clear, matter of fact, fairly unequivocal, reasonable estimate of life span, follow-up with counsellor over following months." (P430)
The current state of knowledge	Low	"Not decisive enough - which can lead to false hopes of a mistaken diagnosis." (P305)
	High	"The support and professional advice I am receiving are as good as it gets. Because the disease develops in different parts of the body and at different rates of progression, getting accurate answers on the future is virtually impossible." (P158)
	Low	"Diagnosis and explanation were quite perfunctory with no explanation to follow-up care. Terminology used was outdated and wrong as to specifics. Expected lifespan estimate was optimistic (7 years)." (P341)

(continued)

SPIKE domains	Rating	Quotes illustrating positive and negative experiences categorized by ratings of neurologists' skills and satisfaction with delivery of diagnosis (high and low ratings).
Current research and therapeutic trials	High	"I was invited to be part of their research and had research MRIs and check-ups at regular intervals, maybe 2 or 3 months." (P94)
The Australian MND Register	Low	"Somewhat confused as appointment with neurologist not explained as an MND Research clinic." (P136)
Diagnosis in writing	High	"After the diagnosis I had contact with a representative from the MND registry and she has been in contact ever since. . . Since being in contact with the MND Registry I have had adequate information sent to me." (P258)
Further information on aspects of MND	High	"Thorough delivery of how diagnosis was reached. Hand written copy of doctor's notes given to me on departure which I found handy to refer back on." (P232)
	Low	"I was not given a diagnosis in writing until I insisted on it. I was not advised to seek a second opinion." (P343)
	High	"After initial diagnosis it was 10 days before neurological tests and confirmation of MND. Neurologist saw me same day at (Name) to discuss all aspects" (P180)
Information about MND association	Low	"I was used as a learning subject for a student. Lack of info about types of MND. I wasn't told what type of MND I had. Here's what he said: 'There's no known cause - no cure - no treatment - no remission - the disease is terminal and can progress rapidly.'" (P343)
Relevant MND internet sites	High	"Neurologist said many times to contact MND Association who will help and provide information." (P296)
Estimate of life expectancy	Low	"We wanted more details about MND help and the Assoc - received no guidance." (P30)
EMOTION (Responding empathically to the feelings of patient/family)	High	"One good thing is that the clinic nurse provided a referral to the MND Association website." (P299)
Diagnosis given with warmth, care and empathy	Low	"My knowledge came from internet. Therefore sought advice of 2nd neurologist. The literature given was inappropriate and horrifying - even 3 years down the line." (P66)
More time allowed to express emotions	High	"I felt that it was very important to receive an honest assessment of my prognosis and life expectancy." (P18)
	Low	"I was totally shocked by my initial diagnosis and the estimate of my possible life expectancy." (P273)
Enough time to have these emotions responded to	High	"He detected it very quickly and was so gentle and caring in the way he told me, I was not upset. I was glad to find a name for all my medical problems." (P264)
	Low	"No positive aspects at all (no help nor care). Had the bedside manner of a brick." (P335)
	High	"Neurologist was clinically thorough but uncaring when providing diagnosis of MND. Gave brochure for information. Told me I had 3 years left to live. Was left alone in city 2 hrs from home after diagnosis." (P85)
	Low	"The neurologist was lovely . . . the appointment was in the afternoon and she spent quite a long time with us we did not feel rushed." (P09)
	High	"I received diagnosis by neurologist and then [she] left room so I could maintain my composure in private. Then [she] came back and gave me time to question her before seeing me again next day." (P54)
	Low	"I felt the neurologist was very insensitive. Especially saying it is 'terminal'. I didn't have enough time to comprehend what I had just been told." (P234)
	High	"Despite knowing a lot, expecting the bad mutation, it was a shock. We didn't have time (or empathy?) to discuss feelings and consequences." (P430)
	Low	"The diagnosis was given very professionally with appropriate empathy and a good understanding of my reactions to it - which were calm." (P152)
	High	"More information and more time would have allowed opportunity for me to be questioned about my feelings and for me to respond accordingly." (P205)

(continued)

Table III. Continued

SPIKE domains	Rating	Quotes illustrating positive and negative experiences categorized by ratings of neurologists' skills and satisfaction with delivery of diagnosis (high and low ratings).
STRATEGY (Making a plan and follow-through) A referral to an MND multidisciplinary clinic	High	"After making contact with specialist neurologist, support and care have been all one could hope for. Local allied health, as well as staff at MND clinic, have provided all the information I have needed. Also kept one step ahead with any equipment required. Local rep of MND Association has also been very supportive. After a shaky start in which I felt a bit bereft of knowledge of where to turn, my care has been first class." (P284)
A referral to the MND association	Low	"Was not offered follow-up from initial neurologist. By internet research we made contact with Professor [Name] and had initial consultation... where diagnosis of ALS was given and support through medication and clinic... contact also with MND Association initiated at this stage." (P341)
A follow-up plan for immediate and long-term support	High	"Yes - in hindsight while original neurologist diagnosed and was compassionate - no f/up plan or support was given - we had to source this ourselves - no idea there was an MND association etc. poorly supported until I contacted MND Association - have been well supported since then." (P30)
Community palliative care	Low High Low	"Needed more [support] at beginning - monthly for first 4-6 months, then longer intervals. Need time to ask all Q's, go through anger/grief, then settle into monitoring and adapting to progress of disease." (P52) "It was just a diagnosis to the neurologist and no emotional support or ongoing guidance was given." (P341) "... put into hospital and the palliative care Dr took over from the neurologist." (P389) "After diagnosis, the neurologist providing 2nd opinion became regular neurologist at MND clinic. Now regularly see palliative specialist Dr - appointments every 3 months." (P201)

(exploring what patient/family are expecting or hoping for) where highly rated neurologists made the effort to ask how much the patient already knew about their condition and how much detail they wished to have; and 4) Strategy (making a plan and follow-through) where satisfied patients appreciated having discussions about referrals to the MND association, to the multidisciplinary clinic, the role of palliative care, the role of community support and a follow-up plan for immediate and long-term support.

It is evident that the longer the patients spent with their neurologists during breaking this bad news, the more they were satisfied with the delivery process and the higher they rated the neurologist's abilities/skills. The median length of the consultation reported in this study was 30 min. However, delivering the diagnosis is a process that requires 45–55 min according to patients who rated the ability of the neurologist 'good to excellent' (Figure 1), or those who were 'satisfied to very satisfied' (Figure 2). This is very similar to the standard outlined in the European guidelines on the clinical management of ALS: Breaking the news – communicating the diagnosis, which states that enough time needs to be available on the part of the physician (at least 45–60 min) (12). Regarding follow-up support, respondents reported a median of seven weeks for the first follow-up visit after diagnosis, while the recommended practice should be within two to four weeks or sooner. Only about 40% of patients reported being referred to an MND association (when all should be referred) and 16% received the diagnosis in writing. Some improvements are clearly needed in these areas for the patients to feel more supported. Comments in Table III highlight the evident relief in accessing better support once patients were connected to the MND associations. Having the diagnosis in writing would help the patient and family communicate the diagnosis to such support organizations.

Other current practices reported in this study that align with the European guidelines include that for 95% of patients the diagnosis was communicated by a consultant neurologist and for 70% of patients the diagnosis was communicated in a stepwise fashion over two or three visits. Also, the majority of patients (96%) reported that the diagnosis was given in a completely private space and had no interruptions while given the diagnosis (91%), and that 70% had a relative present with them when given the diagnosis.

With 29% response rate to the survey, we cannot be certain of the representativeness of this group of the general population of people with MND in Australia, nor does Australia have a comprehensive register that captures all cases in order to make comparisons. However, the profile of respondents seemed to be similar to another comparable study

that used a survey in terms of age, gender and marital status (18). We cannot also be certain if more satisfied or dissatisfied people made the effort to respond. In the McCluskey et al.'s survey (2004) in Philadelphia, USA, where 65% responded ($n=144$), a larger proportion (56%) gave low ratings of their neurologists (18). However this survey was conducted in just one region of the country compared to our national coverage.

Recall of details may not be a considerable issue in this study, as the median time from diagnosis was about 1.5 years where recollections of receiving the diagnosis are possibly still unaffected, in contrast to McCluskey et al., where on average patients were trying to remember details of what happened six years earlier (18).

Because of the anonymous nature of the survey, we cannot tell the number of neurologists involved in these patients' assessments. However, there is a nearly equal and good representation from all five Australian states to give confidence that an adequate number of neurologists are likely to have been involved in these assessments. Nevertheless, it would be valuable to have neurologists also report themselves on their practices (forthcoming article).

This survey is based on one protocol (SPIKES) and we may have obtained different results had we used other protocols. However, the European guidelines support both the positive and negative findings based on the SPIKES protocol (12). Finally, although our survey instrument has not been tested through a formal validation analysis, given that it has stemmed from the SPIKES protocol, it does possess face validity and our findings suggest that most domains within our questionnaire have good internal consistency (Table II).

In conclusion, this study, in its quantitative and qualitative components, has highlighted 'what it takes to make it better' for patients who were receiving the diagnosis, in terms of the neurologist showing more empathy, having longer consultation times and shorter follow-up periods, more referrals to MND associations, and the neurologist sharing more information. These are issues also encountered in other countries and for other life-limiting illnesses and therefore this study is likely to have implications at the international level (22,23). With over one-third of patients dissatisfied with their experience, there is room for improvement in the practice of neurologists. The following comment captures a number of areas needing improvement: "All neurologists need to be sensitive that the way they give the diagnosis will have ongoing impacts for life. 'There is no going back' applies not only to the words themselves but the way in which they are delivered. I appreciate honesty as long as it is sensitively delivered." (P205). Such improvements may be attainable through educational programmes

and the development of best practice protocols with applicability at the international level. Improving the delivery of the MND diagnosis is central to quality care and the benefits would be for both patients/families and the neurologists, as breaking such daunting news is challenging for both groups, the givers and the receivers.

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 people with MND who contributed to this survey. Also, special thanks for the advice of Dr Paul Talman and for the assistance of Kathy Deas with compiling the qualitative data.

Declaration of interest: There are no conflicts of interest with this paper.

References

- Mitchell JD, Borasio GD. Amyotrophic lateral sclerosis. *Lancet*. 2007;369:2031–41.
- Merrilees J, Klapper J, Murphy J, Lomen-Hoerth C, Miller BL. Cognitive and behavioural challenges in caring for patients with frontotemporal dementia and amyotrophic lateral sclerosis. *Amyotroph Lateral Scler*. 2010;11:298–302.
- Mitsumoto H, Rabkin JG. Palliative care for patients with amyotrophic lateral sclerosis: 'prepare for the worst and hope for the best'. *JAMA*. 2007;298:207–16.
- Kiernan MC, Vucic S, Cheah BC, Turner MR, Eisen A, Hardiman O, et al. Amyotrophic lateral sclerosis. *Lancet*. 2011;377:942–55.
- Zoccolella S, Beghi E, Palagano G, Fraddosio A, Guerra V, Samarelli V, et al. Predictors of long survival in amyotrophic lateral sclerosis: a population based study. *J Neurol Sci*. 2008;268:28–32.
- Aoun SM, Connors SL, Priddis L, Breen LJ, Colyer S. Motor neuron disease family carers' experiences of caring, palliative care and bereavement: an exploratory qualitative study. *Palliat Med*. 2012;26:842–50.
- Locock L, Brown JB. 'All in the same boat'? Patient and carer attitudes to peer support and social comparison in motor neuron disease (MND). *Soc Sci Med*. 2010;71:1498–505.
- O'Brien MR, Whitehead B, Jack BA, Mitchell JD. The need for support services for family carers of people with motor neuron disease (MND): views of current and former family caregivers – a qualitative study. *Disabil Rehabil*. 2012;34:247–56.
- Aoun SM, Bentley B, Funk L, Toye C, Grande G, Stajduhar KJ. A 10-year literature review of family caregiving for motor neuron disease: moving from caregiver burden studies to palliative care interventions. *Palliat Med*. 2013;27:437–46.
- Baile WF, Buckman R, Lenzi R, Glober G, Beale EA, Kudelka AP. SPIKES – a Six-Step Protocol for Delivering Bad News: Application to the Patient with Cancer. *Oncologist*. 2000;5:302–11.
- Clayton JM, Hancock KM, Butow PN, Tattersall MH, Currow DC, Adler J, et al. Clinical practice guidelines for communicating prognosis and end-of-life issues with adults in the advanced stages of a life-limiting illness, and their caregivers. *Med J Aust*. 2007;186:S77, S9, S83–108.

12. Andersen PM, Borasio GD, Dengler R, Hardiman O, Kollwe K, Leigh PN, et al. Good practice in the management of amyotrophic lateral sclerosis: clinical guidelines. An evidence based review with good practice points. EALSC Working Group. *Amyotroph Lateral Scler.* 2007; 8:195–213.
13. Miller RG, Jackson CE, Kasarskis EJ, England JD, Forshew D, Johnston W, et al. Practice parameter update: the care of the patient with amyotrophic lateral sclerosis. Multidisciplinary care, symptom management, and cognitive/behavioural impairment (an evidence based review). Report of the Quality Standards Subcommittee of the American Academy of Neurology. *Neurology.* 2009;73:1227–33.
14. MND Association (UK). Best practice guide. Nottingham, UK.: Author*; 2011.
15. Foley G, Timonen V, Hardiman O. Patients' perceptions of services and preferences for care in amyotrophic lateral sclerosis: a review. *Amyotroph Lateral Scler.* 2012;13: 11–24.
16. Chio A, Montuschi A, Cammarosano S, de Mercanti S, Cavallo E, Ilardi A, et al. ALS patients and caregivers communication preferences and information seeking behaviour. *Eur J Neurol.* 2008;15:55–60.
17. Belsh JM, Schiffman PL. The amyotrophic lateral sclerosis (ALS) patient perspective on misdiagnosis and its repercussions. *J Neurol Sci.* 1996;139 (Suppl):110–6.
18. McCluskey L, Casarett D, Siderowf A. Breaking the news: a survey of ALS patients and their caregivers. *Amyotroph Lateral Scler.* 2004;5:131–5.
19. O'Brien MR, Whitehead B, Jack BA, Mitchell JD. 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. *Amyotroph Lateral Scler.* 2011;12:97–104.
20. Malterud K. Qualitative research: standards, challenges, and guidelines. *Lancet.* 2001;358:483–8.
21. Hugel H, Grundy N, Rigby S, Young CA. How does current care practice influence the experience of a new diagnosis of motor neuron disease? A qualitative study of current guidelines-based practice. *Amyotroph Lateral Scler.* 2006;7:161–6.
22. Connolly S, Galvin M, Hardiman O. End-of-life management in patients with amyotrophic lateral sclerosis. *Lancet Neurol.* 2015;14:435–42.
23. Oliver D, Borasio G, Voltz R, Caraceni A, de Visser M, Lorenzi S, et al. The development of a consensus paper on palliative care in neurology – the implications for ALS care. 25th International Symposium on ALS/MND. Poster Communications. Theme 5: Multidisciplinary Care and Quality of Life. *Amyotroph Lateral Scler Frontotemporal Degener.* 2014;15:117–8.