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RESEARCH ARTICLE

Stigma experienced by ALS/PMA patients and their caregivers: a mixed-methods study

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Abstract

Objective: Previous work suggests that stigma negatively impacts quality of life in people living with amyotrophic lateral sclerosis (ALS) and progressive muscular atrophy (PMA). This study aimed to explore experiences of enacted stigma (experienced discrimination) and felt stigma (shame, fear of exclusion) among Dutch ALS/PMA patients and their caregivers. A secondary aim was to assess associated factors of enacted/felt stigma among patients. **Methods:** A two-phase mixed-methods study was conducted, comprising cross-sectional surveys among 193 ALS/PMA patients and 87 caregivers, and semi-structured interviews with 8 ALS/PMA patients and 11 family caregivers. Descriptive and multivariable regression analyses along with qualitative content analysis were used to analyze survey and interview data. **Results:** Survey findings indicate that patients and caregivers experience enacted and felt stigma. Interviews with both patients and caregivers revealed two manifestations of enacted stigma, including social exclusion (e.g. relationship distancing) and stigmatizing attitudes/behaviors displayed by others (e.g. staring), and three manifestations of felt stigma, including alienation (e.g. shame/embarrassment), perceived discrimination (e.g. feeling judged) and anticipated stigma (e.g. fear of exclusion). Patients and caregivers engaged in concealing and resisting responses to stigma. More bulbar symptoms, King's clinical stage, younger age and living without a partner were significantly associated with enacted/felt stigma among patients. **Conclusions:** Our findings reveal a range of perceptions and experiences underlying enacted/felt stigma among ALS/PMA patients and their caregivers that may serve as conversation topics in clinical practice. Future research may shed more light on the determinants as well as the consequences of stigmatizing experiences among patients and caregivers.

Keywords: Amyotrophic lateral sclerosis, stigma, quality of life, mixed methods

Introduction

In absence of a cure, optimizing quality of life is a major focus in the care for those living with amyotrophic lateral sclerosis (ALS) and progressive muscular atrophy (PMA). There are indications that quality of life among ALS/MND patients is among others impacted by experiences of health-related stigma (1,2). Health-related stigma resulting from visible symptoms and the use of assistive technologies and devices (3–6), may lead to social isolation (3,5,7), psychological distress (7,8), and

reluctance to seek support or use assistive devices (3,6), thereby impairing patients' quality of life.

Health-related stigma can be defined in terms of enacted and felt stigma (9). Enacted stigma involves the actual experience of discrimination or social exclusion owing to one's health condition. Felt stigma refers to shame of being deviant and the feeling that discrimination or social exclusion will happen (8). Previous research (1) indicates that although both forms of stigma can be considered determinants of quality of life in people with ALS, felt stigma is a stronger determinant.

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Felt stigma not only impacts ALS/PMA patients, but also potentially impacts their caregivers, referred to as *affiliate stigma*. Affiliate stigma is felt stigma experienced by caregivers due to affiliation with a person with ALS/PMA (10). Despite a paucity of research on stigma among caregivers of ALS/PMA patients, research in other fields suggests caregivers are at risk of experiencing affiliate stigma resulting in increased caregiver burden and diminished quality of life (11–13).

Given the putative adverse impact of stigma on patients' and caregivers' quality of life (1,7), stigma deserves attention as a potential target in the care of people living with ALS. To inform clinical practice, we explored experiences of enacted and felt stigma among patients and caregivers. As a secondary aim, we tested potential associated factors of enacted and felt stigma among patients.

Materials and methods

Study design

A two-part sequential explanatory mixed-methods design was used, comprising (1) cross-sectional surveys among Dutch ALS/PMA patients and caregivers, aimed at exploring (common) experiences of stigma and determining potential associated factors, and (2) semi-structured interviews with Dutch ALS/PMA patients and caregivers, aimed at exploring manifestations of stigma in more depth and identifying possible responses to stigma.

Part 1. Survey

Recruitment. Adults (≥ 18 years) diagnosed with ALS/PMA or caring for a person with ALS/PMA and willing and able to provide informed consent were eligible to take part in the survey. Patients accessed via a research database including all Dutch ALS/PMA patients who have given prior consent for participation in research. A total of 350 eligible patients were selected. Information about the study and an online survey link were distributed among patients through e-mail. Participating patients were asked for permission to contact their primary caregiver to participate in the survey. 127 patients gave permission and provided contact information of their caregiver. Consequently, a total of 127 caregivers were invited to participate in the survey, using a similar email as for patients. The final sample consisted of 193 patients and 87 caregivers (response rates of 55 and 69%, respectively). Participants' characteristics are shown in Table 1.

Data collection. A patient and caregiver survey were created. Both surveys were administered online using Castor's Electronic Data Capture software (www.castoredc.com) and were open for four weeks in fall 2021. Each survey started with

demographic and disease-related questions (e.g. age, diagnosis) followed by questions about stigma. The Stigma Scale for Chronic Illness (SSCI) was used to measure enacted stigma (11 items, e.g. "Because of my illness, some people avoided me") and felt stigma (13 items, e.g. "I felt embarrassed about my speech") among patients (14). Items were rated on a 5-point Likert scale ranging from 0 = never to 4 = always, with higher scores reflecting higher levels of stigma. Stigma among caregivers of people with ALS/PMA was assessed using four items adapted from the Affiliate Stigma Scale (ASS) and eight additional items. Three items measure enacted stigma (e.g. "Other people avoid me because I have a relative with ALS") and nine items measure felt stigma (e.g. "The behavior of my family member with ALS is embarrassing"). Items were scored on a scale from 1 (totally disagree) to 4 (totally agree). Higher scores reflect higher levels of affiliate stigma. Patients also completed the Amyotrophic Lateral Sclerosis Functional Rating Scale – Revised (ALS-FRS-R), to measure functional impairment (12 items scored 0–48), with lower scores indicating greater impairment (15). ALS-FRS-R scores were used to classify patients according to King's clinical staging system for ALS (16).

Analysis. Data were analyzed in SPSS version 25. Descriptive statistics were used to summarize survey data. A per-item frequencies analysis was conducted to indicate what types of stigmatizing experiences are most common. Multivariable regression analyses (method enter) served to explore associated factors of enacted/felt stigma among patients. Data were transformed using a square-root transformation to meet the assumptions of regression analysis. Significance was set at $p < .05$.

Part 2. Interviews

Recruitment. We recruited patients diagnosed with ALS/PMA and current or former caregivers of a person with ALS/PMA aged ≥ 18 years, to participate in an interview. Bereaved caregivers were allowed to participate if the death of the care recipient was no longer than one year ago. We recruited among patients and caregivers who participated in the survey and consented to being approached for participation in an interview. Among survey participants, we purposively invited 20 patients and 12 caregivers with a range of characteristics that may affect manifestations of and responses to stigma (e.g. stigma scores on the SSCI, sex, diagnosis). This resulted in five patients and eight caregivers who took part in an interview. Additionally, three patients and three caregivers were recruited via an advertisement posted on the website and social media of the Dutch ALS patient

Table 1. Demographic and disease-related characteristics of the participants.

| | Patients (N = 193) | Caregivers (N = 87) |
|---|--------------------|---------------------|
| Age, years | | |
| Mean (SD) | 64.6 (10.0) | 63.0 (11.3) |
| Median | 65.0 | 65.0 |
| Range | 28–89 | 28–83 |
| Gender, n (%) ^a | | |
| Male | 140 (72.5) | – |
| Female | 53 (27.5) | – |
| Relationship to patient, n (%) | | |
| Partner | – | 78 (89.7) |
| Son/daughter | – | 2 (2.3) |
| Parent | – | 3 (3.4) |
| Friend | – | 2 (2.3) |
| Other | – | 2 (2.3) |
| Nationality, n (%) ^b | | |
| Dutch | 190 (98.4) | 84 (96.6) |
| Other | 3 (1.6) | 3 (3.4) |
| Living situation, n (%) | | |
| With partner | 119 (61.7) | 64 (73.6) |
| With partner and children | 44 (22.8) | 17 (19.5) |
| Alone with children | 5 (2.6) | 1 (1.1) |
| Alone | 22 (11.4) | 3 (3.4) |
| With others | 3 (1.6) | 2 (2.3) |
| Educational level, n (%) ^c | | |
| Low (primary school, lower vocational education) | 43 (22.3) | 12 (13.8) |
| Intermediate (secondary school, vocational education) | 78 (40.4) | 46 (52.9) |
| High (higher vocational education, university) | 69 (35.8) | 28 (32.2) |
| Work situation, n (%) | | |
| Paid employment | 29 (15.0) | 38 (43.7) |
| No paid employment | 77 (39.9) | 10 (11.5) |
| Retired | 87 (45.1) | 39 (44.8) |
| Diagnosis, n (%) | | |
| ALS | 150 (77.7) | 74 (85.1) |
| PMA | 43 (22.3) | 13 (14.9) |
| Time since diagnosis, n (%) | | |
| <1 year | 47 (24.4) | 21 (24.1) |
| 1–3 years | 76 (39.4) | 42 (48.3) |
| 3–5 years | 31 (16.1) | 9 (10.3) |
| ≥5 years | 39 (20.2) | 15 (17.2) |
| Months as a caregiver, n (%) | | |
| <6 months | – | 16 (18.4) |
| 6 months to 1 year | – | 12 (13.8) |
| 12 months | – | 12 (13.8) |
| >12 months | – | 47 (54.0) |
| Type of onset, n (%) | | |
| Spinal-onset | 149 (77.2) | 68 (78.2) |
| Bulbar-onset | 35 (18.1) | 15 (17.2) |
| Respiratory-onset | 9 (4.7) | 4 (4.6) |
| Functional impairment (ALS-FRS-R) | | |
| Mean (SD) | 31.5 (9.9) | – |
| Median | 33 | – |
| P25%, P75% | 25, 39 | – |
| Range | 1–48 | – |
| Clinical stage, n (%) | | |
| Stage 1 ^d | 31 (16.1) | – |
| Stage 2 ^e | 47 (24.4) | – |
| Stage 3 ^f | 61 (31.6) | – |
| Stage 4 ^g | 54 (28.0) | – |
| Perceived severity of symptoms, n (%) | | |
| Not severe | 25 (13.0) | – |
| Quite severe | 58 (30.1) | – |
| Severe | 63 (32.6) | – |
| More than severe | 20 (10.4) | – |
| Very severe | 27 (14.0) | – |

Note. ALS: amyotrophic lateral sclerosis; ALS-FRS-R: Amyotrophic Lateral Sclerosis Functional Rating Scale – Revised; PMA: progressive spinal muscular atrophy.

^aDue to a human mistake, no information is available on caregivers' gender.

^bOther nationalities include Belgian ($n = 1$), German ($n = 1$) and Italian ($n = 1$).

^cEducational level refers to the highest level of education completed.

^dFunctional involvement of one central nervous system (CNS) region. This group also includes patients who did not report any impairments ($n = 4$).

^eFunctional involvement of two CNS regions.

^fFunctional involvement of three CNS regions.

^gNeed for gastrostomy or noninvasive ventilation.

association. Recruitment continued until inductive thematic data saturation was achieved.

Data collection. From December 2021 to March 2022, interviews were held with 8 ALS/PMA patients and 11 caregivers of whom 10 current caregivers and one former caregiver. The interviews aimed to explore manifestations of stigma in more depth and to identify strategies that patients/caregivers use to cope with stigma. Topics included: social life (e.g. “What is it like to have (a partner with) ALS/PMA in social situations?”), self-image (e.g. “Do you ever feel shame or guilt?”) and self-compassion (e.g. “How do you deal with emotions caused by ALS/PMA?”). Findings in relation to the first two topics are discussed in this paper. Both patients and caregivers were interviewed individually, via telephone or online depending on participants’ preferences, and verbally ($n = 18$) or via written responses ($n = 1$) depending on participants’ ability to speak. Interviews were conducted by MSS, using a semi-structured interview guide including open-ended and probing questions. Interviews lasted on average 46 min (range 27–79 min).

Analysis. Recordings were transcribed verbatim, anonymized, and analyzed in NVivo 12 using conventional and directed qualitative content analysis (17). Based on previous research including but not limited to the field of ALS, a preliminary list of codes was developed (directed approach). After familiarization with the narratives, codes were

applied to the data and modified, supplemented and refined based on the interview data (conventional approach). Conceptualizations of codes were discussed by three team members (MSS, AB, MSK) until consensus was reached. MSS coded all transcripts. Five coded transcripts were checked for consistency by a second researcher (AB). As the data analysis progressed, codes were grouped into overarching themes which all team members agreed upon.

Ethics

This study was approved by the institutional review board of the University Medical Center Utrecht. Digital written informed consent was obtained from all survey respondents and from all patients and caregivers who participated in an interview.

Results

Manifestations of enacted stigma

Survey findings. In the patient sample, enacted stigma scores ranged from 0 to 24, with a median of 4 (P25–P75: 2–8). The most commonly reported experiences of enacted stigma by patients were “others feeling uncomfortable”, “being avoided” and “people staring” (Figure 1). As shown in Figure 2, staring was also experienced by nearly one third of caregivers ($n = 27$, 31.4%).

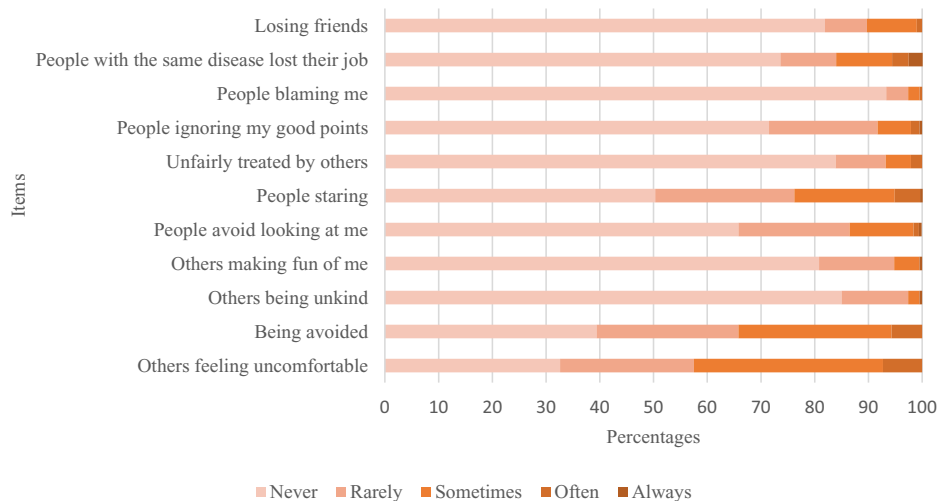


Figure 1. Per-item frequencies for enacted stigma among patients ($N = 193$).

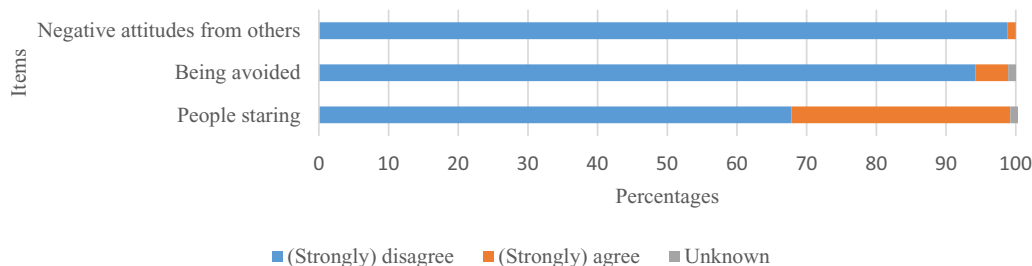


Figure 2. Per-item frequencies for enacted stigma among caregivers ($N = 87$).

Interview findings. Two manifestations of enacted stigma emerged from the interviews: (1) experiences of social exclusion and (2) stigmatizing attitudes and behaviors displayed by others. These themes are presented in Table 2 including sub-themes and illustrative quotes. Experiences of social exclusion were mainly reported by patients and minimally by caregivers and included workplace discrimination, exclusion in social events or gatherings, exclusion in conversations, and relationship distancing. Relationship distancing was experienced by both patients and caregivers who sometimes felt that ALS was a barrier for friends or colleagues to bring a visit to their home. Exclusion in conversations was experienced by patients with impaired speech as well as those using a wheelchair. When being in public, patients with ALS elicit stigmatizing attitudes and behaviors from other people, including avoidance, staring, over-concern or over-attention and patronizing. Patients and caregivers argue that such negative responses can be attributed to visible physical symptoms of ALS, impaired speech and the use of assistive devices such as a wheelchair. Patients and caregivers also experienced being patronized, for example by friends, relatives, colleagues or strangers offering unwanted help or unsolicited advice. They further indicated that ALS sometimes prompts discomfort in other people. Moreover, patients and caregivers sometimes received reactions of incomprehension, not only from their social network but also from healthcare professionals or authorities.

Manifestations of felt stigma

Survey findings. Patients' median score on felt stigma was 9 (score range: 0–51, P25–P75: 4–15). As shown in Figure 3, the most common experiences of felt stigma among patients included “feeling a burden”, “feeling left out” and “feeling different from others”. Feeling left out was also experienced by nearly 20% of caregivers (19.5%) (Figure 4).

Interview findings. The interviews indicated that felt stigma among patients and caregivers manifests itself in three ways, including (1) experiences of alienation, (2) perceived discrimination and (3) anticipated stigma (Table 3). In the domain of alienation, experiences identified were feeling ashamed/embarrassed, different or “other”, inferior, isolated and/or left out. Among patients, these feelings were generally caused by a changing physical appearance, the use of assistive devices (e.g. wheelchair), and when having to ask for help. For their carers, experiences of alienation seem linked to a reduced ability to socially participate. A number of patients indicated feeling a burden to their family or colleagues. On the contrary, some patients and caregivers felt that other people were

incapable of understanding their burden. Both patients and caregivers perceived being not accepted, ignored, judged or watched by others. Anticipated stigma included experiences of anticipated shame and fear of enacted stigma. Some patients anticipated shame and stigmatizing reactions when their disease becomes more visible, e.g. due to worsening of physical symptoms or first-time use of assistive devices. One caregiver feared being devalued and rejected by her colleagues when having to cut hours of work to care for her husband with ALS.

Responses to enacted/felt stigma

Interviews revealed that patients and caregivers engaged in both concealing and resisting responses to stigma (Table 4). Concealing responses involved, among others, social withdrawal. Patients, for instance, reduced or stopped their visits to theaters, restaurants and friends. Furthermore, a former caregiver described how his wife maintained secrecy about the seriousness of her disease toward her friends. Others were able to resist stigma through disclosure, adopting a positive outlook and/or deflecting.

Associated factors of enacted/felt stigma among patients

In an explorative multivariable regression analysis (Table 5), we identified four possible associated factors of enacted stigma among patients, including ALS-FRS-R bulbar score, clinical stage 3 (i.e. functional involvement of three central nervous system (CNS) regions), age and living situation (i.e. with or without a partner). Patients with higher bulbar scores or 3 affected CNS regions, patients under the age of 65 and those living without a partner were found more likely to encounter enacted stigma. Associated factors of felt stigma were highly similar. We found that patients under the age of 65, patients with higher bulbar scores and those with 2–3 affected CNS regions were significantly more likely to experience felt stigma.

Discussion

This study highlights experiences of enacted and felt stigma among ALS/PMA patients and their caregivers. Some of the most common experiences of both patients and caregivers include being stared at (enacted stigma) and feeling left out (felt stigma). In addition, we found that patients with a younger age, living without a partner, having bulbar symptoms, or in an intermediate clinical stage are more prone to enacted and felt stigma.

Both our survey and interview findings support the notion that stigma faced by ALS/PMA patients manifests itself in two ways: enacted and felt stigma. Whereas enacted and felt stigma scores

Table 2. Manifestations of enacted stigma identified from the interviews.

| Themes | Subthemes | Definitions | Example quotes |
|--------------------------------------|--------------------------------|--|---|
| Social exclusion | Workplace discrimination | Experienced discrimination or intolerance in the workplace (e.g. being forced to quit one's job). | "And when I came back from holiday and talked to the company doctor, he literally said 'How can I report you better? You no longer have a future.' I found that so harsh. I have worked very hard year after year, I enjoy my work. It's the only thing you have to hold onto in your life, actually. And, simply because you have a diagnosis, that is stopped and someone else decides about that". (patient) |
| | Relationship distancing | Friends, relatives, neighbors or colleagues avoid/move away from the person with ALS. | "My acquaintances find it difficult. So they don't come by very often. They sometimes phone, but not like they used to". (patient) "Those kinds of conversations don't take place and at a certain point, it just fades". (patient) |
| | Exclusion in conversations | Not being included in conversations (e.g. people talking to caregiver instead of patient). | "In conversations with more than 1 person, I am easily forgotten. People mean well, but are impatient. They easily forget to involve you". (patient) "At first, I was constantly being asked how he [partner with ALS] was doing while he was right there. And I think people do that anyway if someone is in a wheelchair. I said: 'Well, you can ask him yourself. Just ask'". (caregiver) |
| | Exclusion in social events | Being excluded in social events/gatherings (e.g. not being invited to birthday parties). This also includes being invited to events wherein you cannot participate. | "Then you have the team outing, playing volleyball. That is great, but I can't do that". (patient) "If there are birthdays, that they say 'Oh she can't come, can she?'" (patient) |
| Stigmatizing attitudes and responses | Being avoided | People looking away or walking the other way. | "When we walk together, she in the wheelchair and I behind it, you notice the effect of limited greetings and contact". (caregiver) "No verbal responses, but people do react. Looking away, looking the other way". (caregiver) |
| | Staring | People staring at the person with ALS. | "The first couple of times when I was in the wheelchair ... Then you actually get to go somewhere now and then. People stare at you from head to toe". (patient) "When you go into the shop with that [dropped head syndrome], people will look. And if your walking gets worse and worse, people will look at you". (caregiver) |
| | Over-concern or over-attention | Receiving excessive attention or sympathy from others. | "We are sitting there having dinner with friends and then a woman leans over me and says: 'You must be enjoying yourself, aren't you?', but she doesn't say it to the four of us. No, she really leaned over me while saying that". (patient) "People who know us superficially, like the neighbors, they are always a bit careful and 'Are you okay?' and that sort of thing". (caregiver) |
| | Patronizing | People acting over-protective or like they know what you need (e.g. giving unsolicited advice on how to handle the ALS situation, offering unwanted help). | "I'm getting worse and people see that, obviously. And then they kind of want to protect you. And I think: I'm still good enough to take care of myself". (patient) "Now that I reported ill, someone said: 'Good that you're here today and if you manage to do something, that's fantastic. And then I thought: no, you can actually count on me'". (caregiver) |
| | Others feeling uncomfortable | The experience that other people feel uncomfortable being around or talking to you (e.g. being afraid to ask questions). | "I notice that people are wary about asking [about my condition]". (patient) "They may be a bit cautious at times or they don't dare to ask a certain question. While you sense that they are curious". (caregiver) "You don't see a lot of colleagues anymore. With ALS, they hardly dare to come around". (caregiver) |
| | Incomprehension | Extent to which people living with ALS receive reactions of incomprehension from their surroundings, including friends, relatives and acquaintances, but also professionals and authorities. | "I do have that with certain people. That they thought I could do more than I can". (patient) "But also, for example, if you need aids, they sometimes say 'Yes, it will come in 6 weeks'. Then I think 'No, I need it this week'. I sometimes find that there is a lack of understanding in such things. Sometimes you have to explain very clearly what ALS is and why you need it so badly". (patient) "People see what they see and don't realize that speech and walking are the tip of the iceberg for me". (patient) |

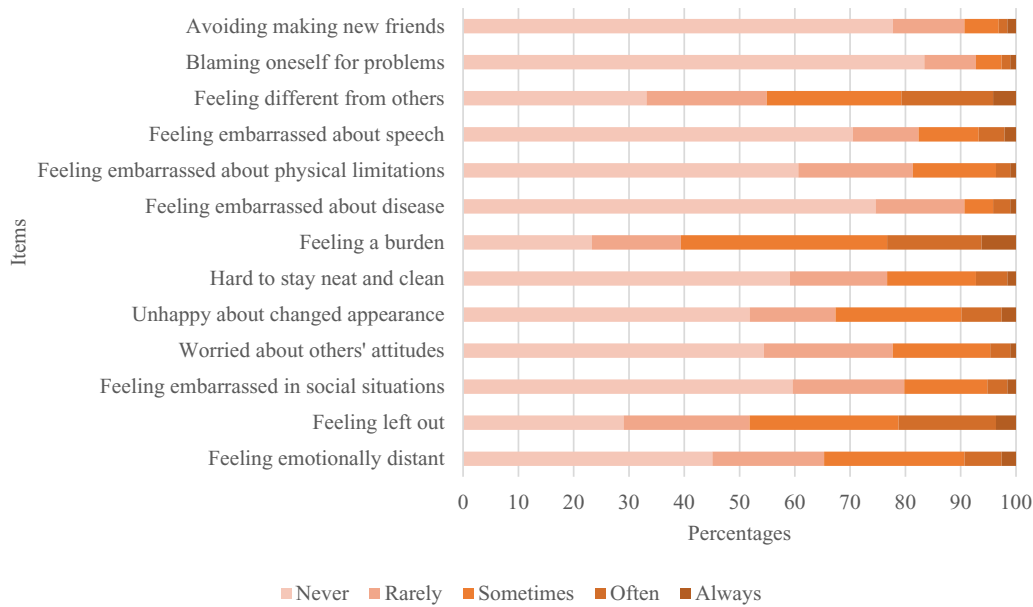


Figure 3. Per-item frequencies for felt stigma among patients (N=193).

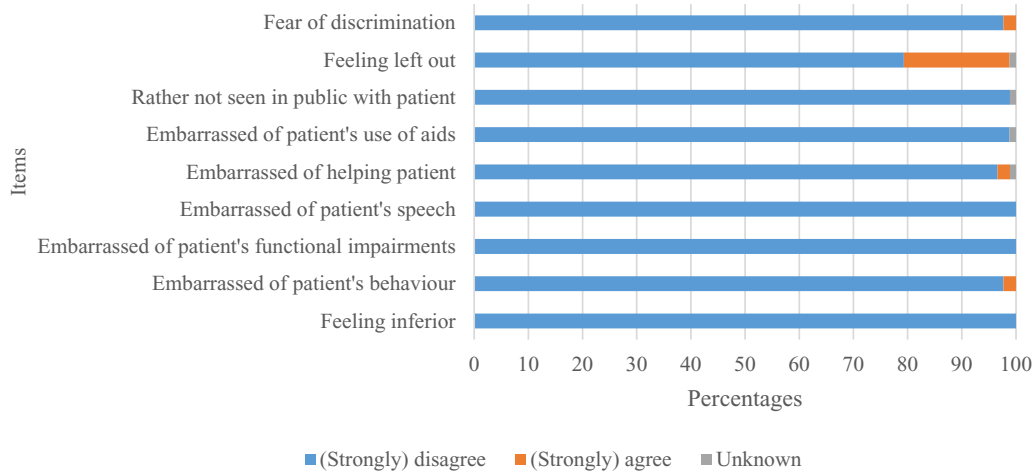


Figure 4. Per-item frequencies for felt stigma among caregivers (N=87).

among patients were comparable to the scores reported in a previous Dutch study (1) among neuromuscular diseases including but not limited to ALS, a study conducted in the United Kingdom (2) yielded considerably higher scores, suggesting cultural differences in experiences of stigma. A recent study (13) showed that levels of affiliate stigma among caregivers of people with Parkinson’s disease (PD) indeed differed across cultures. Such cultural differences may be due to differences in the ways that cultures perceive and respond to illness (18,19). However, differences in stigma across cultures may also be accounted for by differences in the ways healthcare is organized. In the Netherlands, people living with ALS/PMA have access to high quality multidisciplinary care encompassing symptom management and psychological support.

Where experiences of stigma may be culture-specific, such experiences do not seem specific for ALS/PMA. Previous research suggests that people living with cancers, cardiovascular diseases, chronic respiratory diseases and diabetes have many similar experiences of enacted stigma compared to people living with ALS, including workplace discrimination, relationship distancing, exclusion in social events, avoidance and over-attention or over-concern (20–22). This is also true for experiences of felt stigma, such as feeling ashamed/embarrassed, feeling judged or negatively viewed by others, and fear of exclusion (20,22).

Our findings suggest that not only patients, but also caregivers experience stigma, thereby providing more depth to the known experience of caring for a person with ALS/PMA. Similarly to the

Table 3. Manifestations of felt stigma identified from the interviews.

| Themes | Subthemes | Definition | Example quotes (English) |
|--------------------------|--|---|---|
| Alienation | Feeling a burden | Feeling a burden to the people surrounding you. | <p>“I often feel very guilty. I know I can’t help it, but I burden everyone with my problem”. (patient)</p> <p>“I get the feeling from them [family] that I should feel guilty”. (patient)</p> |
| | Feeling ashamed/embarrassed | Experienced feelings of shame/embarrassment associated with the health condition of oneself or one’s partner (e.g. feeling ashamed of physical appearance or speech, having to feed one’s partner). | <p>“It’s still uncomfortable being washed by someone else”. (patient)</p> <p>“At first, I was embarrassed about my speech, but not anymore. I am now embarrassed to eat in front of others, I spill and slurp quite a bit”. (patient)</p> <p>“At first, you sometimes feel embarrassed about situations when you go out to dinner and he spills things or I have to feed him or something”. (caregiver)</p> <p>“At a certain point, she [partner with PMA] found it a bit embarrassing that she was walking badly and always needed support”. (caregiver)</p> |
| | Feeling different | Feeling different or “other” compared to the people (without ALS) surrounding you. | <p>“I do feel different, yes. Of course, I can do less”. (patient)</p> <p>“You can’t really participate, because you’re different, because you’re limited. Everyone walks and talks and you can’t walk, you can’t talk. I don’t want to say it’s isolating, but it’s a different position than before”. (caregiver)</p> |
| | Feeling inferior | Experienced feelings of inferiority associated with the health condition of oneself or one’s partner. | <p>“It is very difficult for me to accept that I can do less and less. It makes me feel inferior”. (patient)</p> <p>“I would have loved to do nice things with my adult children, but I hardly ever do that anymore. So, as a mother of grown-up children, I am worth less and I feel a little less worthy”. (caregiver)</p> |
| | Feeling isolated | Feeling alone/lonely/isolated due to the health condition of oneself or one’s partner. | <p>“You are just isolated in your own body”. (patient)</p> <p>“I think there is a kind of loneliness and I think that is manifesting itself more and more and more, the longer the process lasts”. (caregiver)</p> |
| | Feeling left out | Feeling left out/excluded due to the health condition of oneself or one’s partner. | <p>“My family are getting on with their own things, I feel like I’m out of it”. (patient)</p> <p>“Of course you still hear stories about certain people. On the one hand, that’s nice. On the other hand, it’s also annoying, because you are no longer part of it”. (patient)</p> <p>“I don’t count anymore, in real life”. (caregiver)</p> |
| | Feeling that others are incapable of understanding you | Feeling that the people surrounding you are incapable of understanding your life with ALS. | <p>“Then you also notice that people cannot imagine what it is like for me”. (patient)</p> <p>“There is really no one who actually knows what I experience and feel”. (patient)</p> <p>“I can hardly explain it, what it is like, how difficult it is and what it takes. You have to go through that”. (caregiver)</p> |
| Perceived discrimination | Feeling not accepted | Feeling not accepted by the people surrounding you. | <p>“I think I do accept myself, but I feel that others find that more difficult”. (patient)</p> |
| | Feeling ignored | Feeling ignored/not seen by others (e.g. caregivers feeling they are solely treated as an extension of the patient with ALS). | <p>“I would expect society, but also my immediate environment, to be more involved. Asking more about it, maybe offering help, one way or another. That’s not happening”. (caregiver)</p> <p>“This is obviously a disease that you have together, because as a partner you take over more and more. [...] But, you notice that as far as the outside world is concerned, it is only my husband who is ill, and the illness affects him. And that’s where all the attention goes”. (caregiver)</p> <p>“I’m not just an extension of [partner’s name]. Yet, that is how I am approached, certainly by the medical world”. (caregiver)</p> |
| | Feeling judged | Feeling judged or negatively viewed by others (e.g. feeling that people no longer view you as the person you was, for medical decisions, looking old, people thinking that you’re drunk/dumb). | <p>“And then you also see people looking like: ‘Okay, why can’t she do that herself?’” (patient)</p> <p>“I’ve often noticed that people are startled by the way I talk, think I’m deaf or stupid, and even run away”. (patient)</p> <p>“My husband has been very clear from the start about how far he wants to take it and that quality of life is most important to him and that he doesn’t want to go on to the very end. [...] He talked about euthanasia from the start. Then you also notice that that puts people off or that they have a certain judgment about it”. (caregiver)</p> |

(Continued)

Table 3. (Continued).

| Themes | Subthemes | Definition | Example quotes (English) |
|--------------------|-------------------|--|---|
| | Feeling watched | Sense of being watched by other people. | <p>“When I get up from the chair, I have to think for 2 seconds: ‘okay, what position are my feet in? Can I get right above my legs?’ If that’s the case, then I slide and then at a certain moment 1, 2, 3 and then a kind of swing and then I’m standing. I don’t notice it anymore, but you know, everyone is watching: is he going to make it or not”.</p> <p>(patient)</p> <p>“My wife can’t eat independently. Well, she can, but with a lot of tools. And of course (a) you can’t do that in a restaurant and (b) if you can, it is very noticeable. And I don’t want to say that this has an isolating effect for us, but it is nevertheless a special position that you’re in, as if you are always on a stage. It’s not like that, but it feels a bit like that”. (caregiver)</p> |
| Anticipated stigma | Anticipated shame | Expectation that feelings of shame/ embarrassment will arise under certain circumstances (e.g. when the disease progresses). | <p>“Eating is still going well. But I do notice movement all the time. At some point, I start to tremble. I know that at a certain point I will find that annoying, if it happens in front of others, even if they are my friends so to speak. Suppose you go to a restaurant or somewhere, people will look at you”. (patient)</p> <p>“Yesterday afternoon, I was just sitting working and then he [partner with ALS] suddenly said, ‘I know that I can now, in my wheelchair, for example, pick up the boys from school with you, but I dread seeing the faces of people, that they pity me or that they see how ill I am’”. (caregiver)</p> |
| | Fear of exclusion | Fear of being excluded or treated differently due to the health condition of oneself or one’s partner. | <p>“You know, I really don’t want to be written off. [...] Like someone who just sits at home. You always want to carry on being someone”. (caregiver)</p> |

Table 4. Responses to stigma identified from the interviews.

| Themes | Subthemes | Definition | Example quotes |
|----------------------|-------------------|--|---|
| Concealing responses | Social withdrawal | Withdrawal from or avoidance of social interactions/situations (e.g. visiting friends, birthday parties, going to a restaurant or theater), due to (anticipated) experiences of stigma (e.g. feeling a burden/ embarrassment). | <p>“I find it embarrassing that I can’t get up out of a normal chair. And if I fall, then I need help, to get up. I find that frustrating. Of course, this means you’re going to avoid doing things. I no longer go to the theater, I no longer go to the opera, I no longer go out by car to visit things”. (patient)</p> <p>“That was also a bit of her embarrassment, to not have too much social interactions anymore as she thought she looked really bad at the end”. (caregiver)</p> |
| | Secrecy | Maintaining secrecy about the health condition of oneself or one’s partner. | <p>“My wife, who did have a positive attitude, never actually told her friends, even her closest friends, how bad her condition really was”. (caregiver)</p> |
| Resisting responses | Deflecting | Rebuffing other people’s (negative) attitudes or responses as irrelevant. | <p>“That other people look at me, or whatever, I couldn’t care less”. (patient)</p> <p>“I don’t feel uncomfortable because I’m with him. I sort of think, you have every right to be there and I’m not going to hide or anything”. (caregiver)</p> |
| | Positive outlook | Focusing on the positives rather than on the negatives. | <p>“Of course, there are days when I’m angry, and all I can do is cry all day. But that’s once in a while. I wake up happy every day and try to look at the positives”. (patient)</p> <p>“I still see the positive sides of [partner’s name] more than the ill sides”. (caregiver)</p> |
| | Disclosure | Opening up about ALS and its limitations to other people (e.g. family, friends, relatives, neighbors, colleagues). | <p>“Whether they ask what I still can do with my hands or ask if, so to speak, I can still poop. I don’t care what. They are allowed to know everything about me. I don’t just tell them everything myself, but I answer every question openly”. (patient)</p> <p>“I think you can only expect understanding from people if you are open about what you are confronted with”. (caregiver)</p> |

Table 5. Outcomes of multivariable regression models assessing associated factors of enacted and felt stigma among patients ($N=193$).

| | <i>B</i> | <i>SE B</i> | β | <i>t</i> | <i>p</i> -Value | R^2 |
|-------------------------------|----------|-------------|---------|----------|-----------------|-------|
| SSCI – enacted stigma | | | | | | |
| Constant | 3.91 | 0.66 | | 5.98 | .000* | 0.22 |
| ALS-FRS-R – Bulbar score | 0.07 | 0.03 | 0.17 | 2.35 | .020* | |
| Clinical stage 2 ^a | 0.09 | 0.26 | 0.03 | 0.34 | .731 | |
| Clinical stage 3 ^b | 0.58 | 0.27 | 0.22 | 2.18 | .031* | |
| Clinical stage 4 ^c | 0.34 | 0.28 | 0.12 | 1.20 | .230 | |
| Gender | 0.31 | 0.19 | 0.11 | 1.68 | .095 | |
| Age | -0.03 | 0.01 | -0.28 | -4.00 | .000* | |
| Partner | -0.48 | 0.22 | -0.14 | -2.15 | .033* | |
| Work | -0.35 | 0.25 | -0.10 | -1.39 | .165 | |
| SSCI – felt stigma | | | | | | |
| Constant | 4.13 | 0.82 | | 5.07 | .000* | 0.19 |
| ALS-FRS-R – Bulbar score | 0.08 | 0.03 | 0.18 | 2.42 | .016* | |
| Clinical stage 2 ^a | 0.75 | 0.33 | 0.21 | 2.29 | .023* | |
| Clinical stage 3 ^b | 1.04 | 0.33 | 0.32 | 3.14 | .002* | |
| Clinical stage 4 ^c | 0.67 | 0.35 | 0.20 | 1.92 | .057 | |
| Gender | 0.28 | 0.23 | 0.08 | 1.22 | .224 | |
| Age | -0.04 | 0.01 | -0.24 | -3.39 | .001* | |
| Partner | -0.07 | 0.28 | -0.02 | -.26 | .797 | |
| Work | -0.35 | 0.31 | -0.08 | -1.14 | .256 | |

Note. SSCI scores were transformed using square-root transformation.

ALS-FRS-R: Amyotrophic Lateral Sclerosis Functional Rating Scale – Revised; SSCI: Stigma Scale for Chronic Illness.

* $p < .05$.

^aFunctional involvement of two CNS regions.

^bFunctional involvement of three CNS regions.

^cNeed for gastrostomy or noninvasive ventilation.

patients they care for, caregivers seem to experience both enacted stigma (e.g. relationship distancing, patronizing) and felt stigma (e.g. shame/embarrassment, isolation). While there is a paucity of research on stigma among ALS caregivers, research in other neurodegenerative diseases supports our finding that caregivers are at risk of experiencing affiliate stigma (11–13,23,24).

In the light of previous research suggesting that stigma in neurodegenerative diseases including ALS may adversely impact patients' quality of life (1,2,6) and caregivers' burden (3,11,12), it seems important to address stigma in healthcare. To be able to do so, it is important to understand the factors affecting enacted and felt stigma. In our study, bulbar symptoms were identified as a potential determinant of both enacted and felt stigma, thereby supporting previous research among MND patients (5,25). The extent to which the patient is physically impaired also seems to matter. When 2 or 3 CNS regions are affected, people seem more susceptible to stigma. This may be accounted for by increased visibility of the disease. In our interviews, many patients linked experiences of stigma to physical manifestations of the disease as well as the use of assistive devices which make the disease visible. Consistent with our findings, visible physical symptoms including tremors, rigidity and dysphagia have been associated with experiences of stigmatization

in PD (13,26). Furthermore, in a recent review (20), the visibility of physical symptoms was identified as one of the main causes of stigma among a number of chronic diseases.

Practice implications

Of those patients and caregivers who participated in an interview, many seem to have experienced some forms of stigma yet they seem to cope well with it. Consistent with the literature, our interview findings reveal several coping strategies, including disclosure (i.e. opening up to others about one's disease and limitations) and deflecting (i.e. rebuffing other people's attitudes or responses as irrelevant), which may be applied to clinical practice/interventions with patients and caregivers. Although not apparent in our interviews, other coping skills may contribute to reducing or overcoming stigma as well, for instance socializing with other people living with ALS and increasing knowledge about ALS (20), both of which can be well integrated in ALS/MND multidisciplinary clinic structure.

ALS care professionals may play an important role in supporting and promoting such stigma-resisting strategies among patients and caregivers through creating awareness of stigmatizing experiences that patients and caregivers may encounter due to ALS and discussing such experiences. Our

findings highlight a range of perceptions and experiences underlying stigma that may serve as conversation topics for e.g. rehabilitation physicians, psychologists and social workers. Professionals should pay particular attention to patients who are younger than 65 years, patients living without a partner and those with bulbar symptoms and/or 2–3 affected CNS regions as they are more likely to experience enacted/felt stigma.

Our survey and interview findings suggest that both patients and caregivers have experiences of stigma. As the disease progresses, patients' and caregivers' lives tend to become increasingly intertwined (27), including stigmatizing social experiences. This underscores the importance of involving caregivers in the dialogue about stigma rather than only the patient.

While felt stigma may be modifiable through intervention of healthcare professionals, enacted stigma may be difficult to modify. Tackling enacted stigma, which is often the result from ignorance and negative stereotypes prevalent in society, does not require action from professionals per se, but rather requires society to undergo some changes. Public awareness campaigns and programs promoting inclusiveness may contribute to more understanding and reduce stigmatizing attitudes, beliefs and behaviors in the general public.

Strengths and limitations

This study adds to the literature in multiple ways: (1) in describing experiences of stigma, we addressed both the patient and caregiver perspective; and (2) we used both quantitative and qualitative methods to empirically support the commonly used conceptualization of stigma in enacted and felt stigma. However, there were also some limitations to our study. First, stigma scores may have been underestimated. During the study, everyone was socially more or less isolated due to COVID, hence respondents may have felt less different and left out. Second, differentiation into low, moderate and high levels of stigma was not possible due to lacking cutoff values. Third, we were only able to include one ALS-patient with speech difficulties in the interviews, while previous research (3–6) suggests that impaired speech is a determinant of stigma among patients living with motor neuron disease, which was confirmed by our data. Fourth, there was no validated measure available suitable for self-assessing enacted and felt stigma among caregivers of ALS/PMA patients. Finally, in addition to the demographic and disease-related characteristics included in our study, there are a lot of psychological factors that may affect stigma, such as coping, self-esteem, and depression (26,28). These should be taken into account in future research on determinants of stigma.

Conclusions

Our findings suggest that ALS/PMA patients and their caregivers have experiences of both enacted and felt stigma. Whereas tackling enacted stigma requires a change in society as a whole, ALS care professionals could take an active role in minimizing felt stigma, among others through teaching patients and caregivers resisting (coping) responses to stigma. Future research may shed more light on the determinants as well as the consequences of stigmatizing experiences among patients and caregivers.

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Data availability statement

The data that support the findings of this study are available from the corresponding author, MSS, upon reasonable request.

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