The family experience of living with a person with amyotrophic lateral sclerosis: A qualitative study

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Living with a person with amyotrophic lateral sclerosis (ALS) is a complex and difficult experience. Most research involves only the primary caregiver and uses a quantitative approach. The aim of this study was to explore the experience of family members who live with ALS patients until their death. In-depth, semi-structured interviews were conducted with 13 family members of ALS patients now deceased. Transcripts were analysed using interpretative phenomenological analysis. Three main themes were identified: “Meaning of ALS,” including the peculiarity of ALS and its comparison with other illnesses, the explanation of ALS, emotions, coping strategies, personal change and difficult choices; “Family relationships,” including centripetal vs. centrifugal forces, role changes, ALS as a family disease, ALS as a family solution, openness towards the outside world; and “Healthcare context,” including access to services, information and humanization. One finding was that families of a person with ALS need more supportive interaction and information during the patients’ illness and their end-of-life. This study is an invitation to understand families’ experience and subsequently help them to find new ways to cope with the situation.

Keywords: Amyotrophic lateral sclerosis; Caregiver; End-of-life; Family; Qualitative methods.

Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disease with a sudden onset, a rapid progression, a profile of complex disabilities and fatal consequences. People with ALS experience progressive muscle weakness, becoming progressively immobile, and develop impaired speech and respiration problems. In the late stages of the disease, progressive paralysis can result in a “locked-in” state in which only residual muscular movement is possible, but awareness usually remains unchanged. Despite an average life expectancy of about 3 years after symptom onset, the progression of the disease is unpredictable, with 10% of patients living more than 10 years (Andrews, 2009).

As the disease progresses, ALS patients require assistance with mobility, eating, dressing and nursing care; assistance is often provided by a family member, usually the patient’s partner who assumes the role of primary caregiver. Sometimes, this person spends more than 11 hours per day with the patient (Pagnini, 2013), sometimes also taking advantage of paid assistance.

The progressive nature of the disease accentuates the dependence of the patient upon the primary caregiver, but conclusions drawn from the evidence about the relationship between the caregiver’s psychological status and patient disability are controversial. Some studies have shown that caregiver burden and depression are positively related with ALS patients’ physical disability (Pagnini et al., 2010) and increase over time (Gauthier et al., 2007). In contrast, recent studies have shown a maintenance of, or even an increase in, quality of life (Osloss, Markhede, Strang, & Persson, 2010) and lessening of depression on the part of the caregiver (Rabin, Albert, Rowland, & Mitsumoto, 2009). Moreover, caregivers who present lower levels of quality of life are not always those who have to look after the most physically or psychologically impaired patients (Lo Coco et al., 2005). Finally, social support, finding positive meaning in caregiving, and the quality of the relationship with the ill partner have significant effects on carers’ adjustment (Atkins, Brown, Leigh, & Goldstein, 2010; Chio, Gauthier, Calvo, Ghiglione, © 2014 International Union of Psychological Science
& Mutani, 2005; Goldstein, Atkins, Landau, Brown, & Leigh, 2006; Rabkin et al., 2009).

Most of the research on ALS follows a quantitative approach, and a more in depth understanding of patients’ and caregivers’ experience is required (Aoun, Connors, Priddis, Breen, & Colyer, 2011; Palmieri et al., 2010; Wicks et al., 2007). In one qualitative study (Aoun et al., 2011) family caring was described as a demanding and overwhelming task that is characterised by a series of losses accompanied by hopelessness, a role change from spouse to nurse, difficulty in accepting the diagnosis, the absence of compassion on the part of medical professionals with serious emotional consequences for the caregivers, and difficulty in obtaining timely palliative care.

Another study (Oyebode, Smith, & Morrison, 2013) involved partners of ALS patients and highlighted that they had concerns for patient safety, had social restrictions, were continually tired, struggled with anger and frustration, lost intimacy, and were uncertain about the future. However, the caregivers also attempted to adjust to the situation, trying to be strong, retaining a sense of normality, appreciating the provision of specialist services, adopting a problem-solving approach, living day to day, and trying to remain positive and to inhibit signs of grief in order to appear strong.

Finally, two studies (Baxter et al., 2013; Kühnlein et al., 2008) focused on caregivers’ perspectives on end-of-life and the use of non-invasive ventilation (NIV). Baxter et al. (2013) identified a range of potential barriers to the use of NIV including: adverse impressions of the technology; sleep disturbance; the sensation of pressure and pulsing; dry mouth; and mask design issues. Patients and carers perceived benefits related to: increased energy; improved sleeping; enhanced carer well-being; improved breathing and increased speech clarity. The study highlighted the importance of patient perceptions of gains as a factor in their NIV usage decisions.

NIV is known to improve quality of life and to prolong survival in ALS patients. However, little is known about the circumstances of dying in ventilated ALS patients. In the light of the debate on legalising euthanasia, Kühnlein et al. (2008) provided empirical data about the process of dying in these patients in Germany, where euthanasia is illegal. In contrast to the Netherlands, where 20% of terminal ALS patients die from physician-assisted suicide (PAS) or euthanasia, only a small minority of the German patients who participated in the study thought about PAS. Most of these patients died peacefully at home from carbon dioxide narcosis, but choking was described in some patients with bulbar-onset ALS, a disease variant with poorer prognosis that affects the muscles of the bulbar region and causes dysphagia, dysarthria and respiratory disorders (Simmons, 2005).

Our study was conducted in Italy (where euthanasia is illegal) and our aim was to explore the experience of family members who have lived alongside ALS patients until their death. Previous research has focused exclusively on family primary caregivers, whereas we have also involved other family members because we did not want to focus only on the caring experience. We were more interested in how a person’s life may change if that person’s loved one contracts ALS, how subsequently the relationship between them and the whole of family life changed, and how the adjustment to this situation and to the patient’s death may vary according to the different meanings family members attribute to ALS.

**METHODS**

**Participants**

We recruited participants among respondents to a message published on a website (www.slaitalia.it) that includes a forum that addresses issues related to ALS. Respondents sent an email to the researcher and were subsequently contacted by telephone. They were provided with details of the study and asked for their initial consent to be interviewed. This was followed up with the provision of written consent prior to the interview. Anonymity was assured and the location of the interview was negotiated with the participants. Interviews were conducted at the participants’ home or in a quiet place chosen by the participant (e.g., an unfrequented coffee bar).

The data collection was based on both principles of saturation, the point where variation ceases, usually when the number of interviews reaches around 15 (±10) (Kvale, 2007), and with regard to the maximum number of people that interpretative phenomenological analysis (IPA) recommends (Smith, Flowers, & Larkin, 2009).

The final number of participants was not predetermined. Our purpose was to select participants who wanted to share their experience of living with an ALS patient, regardless of the type of relationship, and the possibility of being the main caregiver or not. We looked for the maximum variability of the family roles in order to explore and compare the experiences of family members who had a different type of relationship with the patient. As respondents were all family members of an ALS patient now deceased, we decided to focus on how this experience was told after the patient’s death. We took the opportunity to explore family experience until the patient death.

The total number of participants was 13 (10 females and three males) with an average age of 44 years (minimum 24, maximum 64), all living in Northern Italy. Two were sons, six daughters, one husband, two wives, one girlfriend and one sister of the patient. Four of the 13 participants were also the patient’s primary caregiver with regard to the patient. In all cases, the patient’s death dated back between six months and five years before the interview.
Data collection

We conducted semi-structured interviews (Kvale, 2007) to obtain rich descriptive information about the phenomenon, and to allow participants to choose how and what to tell us. We adopted a curious and facilitative, rather than a challenging and interrogative, stance.

At the beginning of the interview participants were encouraged to speak freely about their experiences. Next, we asked questions to analyse topics relevant to our goal, such as illness meaning, illness explanation, diagnosis period, adjustment, illness impact on the family, emotions, social environment and future perspectives. The interviews lasted from 40 minutes to 180 minutes, were audio-taped, and transcribed verbatim.

Data analysis

Interview transcripts were analysed using IPA. The aim of this approach is to understand the subjective experience of participants, by identifying recurrent themes in their narratives. Following the guidelines identified by Smith et al. (2009), we proceeded to the analysis of the transcripts using a bottom-up procedure, whereby the researcher generates codes from the data and not from a pre-existing theory, and including a series of recursive steps. The first step involved a repeated reading of the transcripts to become familiar with the participants’ narratives and to annotate what was significant. In the second step, recurrent themes were identified and linked to quotes that expressed the essence of their contents. Next, themes were clustered together into super-ordinate themes. Finally, we individually re-read all interviews to verify whether the identified themes were recognisable in the transcripts and to ensure that all salient themes had been found. Disagreements between our individual interpretations were resolved by discussion.

In accordance with the quality criteria for IPA (Smith et al., 2009), coherence and transparency, were achieved by reporting how interviews were conducted and analysed, and using thick quotes from the interviews. The depth and breadth of the interviews enabled us to claim a comprehensive, authentic understanding of the experiences of the participants. Moreover, an independent auditor reviewed and verified the consistency of this research, and field notes, interview summaries, analysis notes and process notes were preserved for examination and to determine potential bias.

RESULTS

We identified three thematic areas: “Meaning of ALS,” “Family relationships” and “Health care context.” Each thematic area included different themes that describe the participants’ experience.

The meaning of ALS

The thematic area of the meaning of ALS involved seven themes: the peculiarity of ALS and its comparison with other illnesses, the explanation of ALS, emotions, coping strategies, personal change and difficult choices.

ALS was described as a thief of identity and memories because it prevents the caregiver from recognising his/her loved one and make him/her perceive the patient as a stranger, and forget how the patient was before becoming ill.

You are lucid but you’re immobile. ALS doesn’t leave you even the chance to cry, to scream, to vent, to go mad. (Participant 4, daughter)

He became a person who had nothing to do with what he had been before. (Participant 12, girlfriend)

The thing that made me get more angry is that it’s a disease that takes away your beautiful memories. I couldn’t remember his [the patient’s] laugh, voice […] I liked when he yawned because he had the same sound as when he was still healthy. (Participant 3, daughter)

The drama of ALS was compared with other serious illnesses. ALS does not leave any hope, it is a death sentence. It is inconceivable, not only because it is extremely cruel, but also because it throws the family into total uncertainty, since it is not possible to predict in advance what will happen in the course of the disease. Having no logical sense, it is something incommunicable and, at the same time, unacceptable. It strips off everything except lucidity, thus forcing the patient to assist with the horror of his/her decay. So ALS results in being the worst of all imaginable diseases.

I hoped he had cancer, because so at least we could have tried something. (Participant 10, daughter)

It [living with an ALS patient] was living outside any logic. (Participant 2, husband)

Although ALS aetiology is still unknown, the family member, groping in the dark, constantly and obsessively wondered about the possible causes of the disease.

I had a kind of intellectual despair because I couldn’t believe that there was something so serious that had no hint of a way out. (Participant 6, sister)

After my father’s death, I dreamt about him finally telling me the real causes of ALS. (Participant 3, daughter)

The illness, with its ruthlessness, stirred up in the family member a flurry of negative emotions: a mixture of despair, sadness, grief, helplessness, anger and aggression, fear, and exhaustion, which persist until the patient’s death: “It’s as if a person is slipping into a well and you cannot hold her back.” (Participant 7, daughter)
We identified five ways in which the respondents coped with ALS. Most of the people (eight participants) actively faced the illness by trying to find a concrete solution (e.g., alternative methods to cure the illness or ploys to solve difficult situations and satisfy patient's needs and desires such as drinking a beer). Other strategies, sometimes combined with the first one, were: drawing strength from the patient, maintaining a positive attitude, expressing emotions and avoiding emotions.

I am the one who is strong, who faces things, who grasps the nettle. Hiding is useless. (Participant 10, daughter)

He [the patient] gave me the strength to face this illness. [...] The patient can change your perception of the illness. (Participant 3, daughter)

Thinking positively gives you the energy to look for concrete solutions. (Participant 11, son)

We have always cried together. [...] Because I couldn’t keep my despair inside. [...] We have faced the disease with such despair, but with much sincerity. (Participant 13, wife)

I reasoned like a stranger, like a nurse. [...] Since my husband got sick I have no longer been able to cry, [...] to have any kind of feelings. (Participant 1, wife)

Illness radically changed the meaning that participants attributed to their lives. Principles and values guiding action, personal identity and weight given to the events changed, participants began to appreciate little things and fully live their lives. They felt lucky and privileged to be healthy, demonstrated increased awareness, became more human, sociable, mature and stronger and, at the same time, more catastrophic.

I became enriched because such an experience helps you to understand what are the important things of life. (Participant 5, daughter)

Participants also referred to the existence of the difficult choices and crossroads that ALS continually put them in front of, dealing with NIV, euthanasia, or family management.

The will of my husband was not to do a tracheotomy, not to live as a vegetable. But, in our case, the presence of a child, who needed a father figure [...] I thought that a father figure, even in a bed, who didn’t speak, was always better than having a dead father. (Participant 1, wife)

The disease involved disruptions in family dynamics in terms of centripetal forces that make family members move closer together (in the case of ten families), or centrifugal forces that make family members move away from each other (in the case of three families).

We always had a quite united family. And the disease joined us more. (Participant 13, wife)

Busy in following the illness of my wife?! I didn’t know anything. Others took care of her [the patient ...] Even my children were with her as little as possible. (Participant 2, husband)

The illness also triggered role changes: one patient became a sort of baby and the interviewed family member became a sort of parent.

It’s like having a baby. Except that when a baby grows, you go on seeing improvements in small steps, but in this case it is totally the opposite. (Participant 9, daughter)

ALS is perceived as a family illness as if it was contagious: the patient’s sorrow also affected those around him/her. In one case, ALS represented a solution for ensuring the homeostasis of the family system. ALS allowed the family to stay together because it prevented the husband from communicating openly to his wife that he wanted to terminate their relationship.

I felt sick too. I felt like I could not walk, breathe, swallow. [...] As if the illness was contagious [...] It’s a family disease. [...] It’s a disease that devastates the entire family, not only the patient himself. (Participant 7, daughter)

I was able to survive the period of the illness because I thought that the disease saved me from having to confess to my wife that I didn’t love her anymore. (Participant 2, husband)

In two cases, ALS produced an unexpected openness, availability and permeability towards the outside context:

My parents realised that they had to open to the outside world, because opening to the outside world doesn’t mean suffering damage, but growing rich. [...] The disease is an experience that gives you a challenge in relation to others, because you need the others. (Participant 11, son)

**Family relationships**

The thematic area relating to family included five sub-themes: centripetal vs. centrifugal forces, family role changes, ALS as a family illness, ALS as a family solution, and openness towards the outside world.
difficulty of accessing services and finding information about the illness. This led families to learn on their own through their direct experience and via the Internet.

It is said that a lot of aid is available for ALS sufferers. But I didn’t receive anything. Anything. (Participant 5, daughter) They [health care professionals] gave me no information, no one explained to me what would I should have done, no one explained me what was the duration of the disease, what would happen. (Participant 1, wife)

If I had a problem, I never called the hospital. I connected to the internet, looking at what happened to other families. (Participant 3, daughter)

I learned on the internet, not by talking to physicians, all the things. (Participant 7, daughter)

Family members complained of a lack of humanity and empathy on the part of physicians, an aspect that was perceived from the very beginning. The communication of the diagnosis was brutal and cold, or careless and casual, even by letter. A physician who goes beyond medical information and strives to provide knowledge, tips and tricks, was considered warmer and more humane. The inadequacy of the social-health context induced a strong feeling of abandonment and loneliness on the part of the families that the Internet helped them to face.

The doctor was silent, didn’t say anything. “Doctor, does he [the patient] have it or doesn’t he?” The answer was: “Madam, what do you want me to say, that he has it or that he doesn’t have it?” I remained feeling stupid, because that wasn’t the right behaviour. I would have smashed his face. But when you’re in these situations, you are in awe of doctors. I was shocked by his reaction. (Participant 1, wife)

We knew the diagnosis through the remarkable idiocy of a nurse who put her clinic folder on the legs of my wife. She placed it there at random so that she had her hands free to push the wheelchair. We opened it and read “suspected ALS.” (Participant 2, husband)

We felt completely alone, abandoned by the institutions, by doctors, by everyone. (Participant 4, daughter)

You enter a forum and see the solidarity among the people there […] luckily there is the internet! […] You log in, you read the others’ experiences, you write, they respond, they embrace you virtually. (Participant 3, daughter)

DISCUSSION

The participants’ narrations of their experiences as family members who lived with an ALS patient revolved around the meaning of ALS, family relationships and the healthcare context.

Some of the themes highlighted in our study are in line with the findings of previous studies that focused on caregivers’ experiences, such as total devotion to patient caring, changes in relationship between the patient and family member, feelings of anger and frustration, uncertainty about the future, efforts to be or appear strong, adopting a problem-solving approach, trying to be positive, the avoidance of emotions and the de-humanization of healthcare professionals (Aoun et al., 2011; Cipolletta, Marchesin & Benini, 2013; Cipolletta & Oprandi, 2014; Oyebode et al., 2013).

The participants in our study highlighted that a peculiarity of ALS, if compared with other illnesses, is that it represents a death sentence. In the Italian context euthanasia is illegal, therefore, patients and their families cannot choose when and how to end a fatal and progressive impairment due to the illness; they can only choose whether and when to accept NIV. Our respondents did not delve too deeply into this topic, probably because the patient had already died. Therefore, there was no further decision to take. Moreover, their memories were more focused on the experience of assistance in which they were involved so closely. They felt alone in this experience, especially with regard to aid from healthcare services.

We found resentment and recrimination towards the healthcare system because it does not provide enough information and ease of access to services for ALS patients. A previous study (O’Brien, Whitehead, Murphy, Mitchell, & Jack, 2012) underlined that patients also do not access services because families try to retain control and maintain normality within the home, and are not aware of service entitlement. In the current study, and supported by some previous research (Bolmsjö & Hermerén, 2001; Brown, 2003; Oyebode et al., 2013), participants worked to control their feelings to give an impression of strength by focusing on the immediate and the practical, and searching for information and solutions on their own. These families also experienced centripetal forces that brought them closer (Rolland, 1987). This result is in line with previous studies, which have shown high levels of cohesion and adaptability in families coping with ALS (Tramonti, Barsanti, Bongioanni, Bogliolo, & Rossi, 2014). The strengthening of family relationships may represent an adaptive solution to front a progressive and impairing disease like ALS.

On the other hand, we found centrifugal processes that resulted in greater distance between family members. These forces were more frequent in family members who also avoided emotions. Avoidance has been identified as characterising caregivers of patients in a vegetative state who experience high levels of anxiety, depression, family strain and prolonged grief (Cipolletta, Bastianelli, & Gius, 2014). Avoiding emotions may favour somatization (Cipolletta, Consolaro & Horvath, 2013; Cipolletta et al.,
and may prevent people from recognizing and caring about some illnesses (Cipolletta, Beccarello, & Galan, 2012).

The main limitation of our study was that we involved only one person per family, thus preventing us from understanding the experience of the other family members. Moreover, our study participants were the partners, children and sisters of patients with ALS, thereby preventing us from gaining in-depth knowledge of how the type of relationship with the patient may impact the experience of another family member. For future research, it would be interesting to take into account the experience of all members of each family system. Furthermore, it would be interesting to explore change in family roles and structure in a longitudinal study. The experience of family members when the patient is still alive might also be explored. Finally, other specific measures might be used in order to consider some of the specific dimensions of family relationships, such as the distribution of dependences (Cipolletta, Shams, Tonello & Pruneddu, 2013).

Our study was an attempt to combine the different dimensions identified in previous studies, in order to provide a comprehensive understanding of an ALS patient’s family experience. We found that this experience is the result of an interweaving of different aspects dealing with more strictly psychological dimensions, such as changes in family relationships or the meaning of ALS as a life threatening illness, but also social dimensions, such as the illegality of euthanasia and the de-humanization of the healthcare system.

Some practical suggestions for clinical intervention may be derived from these results. Previous studies have indicated that caregivers influence the physical and mental status of patients (Chiò et al., 2005; Cipolletta, Marchesin et al., 2013). Consequently, support interventions aimed at caregivers may have indirect beneficial effects on patients’ health (Atkins et al., 2010). Caregivers should take care not only of the patient, but also of themselves, in order to offer adequate support to their loved ones (Rabkin & Albert, 2009). Patient and caregiver should offer mutual support: caregivers should support the patient by taking care of them, and patients should support caregivers by appreciating and encouraging them in their work (Chiò et al., 2005). Our attempt to understand the complex relationship between patient and family may be useful in terms of conceiving psychological support that takes into account the role of illness within the family. Our study may also allow physicians to go further with regard to the medical dimension of ALS and consider its implications for the family system, thus encouraging a humanization of medical care.

REFERENCES


