

Reflections of family caregivers and health professionals on the everyday challenges of caring for persons with amyotrophic lateral sclerosis and cognitive impairments: a qualitative study

Lene Klem Olesen , Karen la Cour, Heidi With and Charlotte Handberg 

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Abstract

Aims and objectives: To explore reflections of family caregivers and health professionals regarding the challenges involved in caring for persons with amyotrophic lateral sclerosis and cognitive and/or behavioral impairments (PALS/CIs).

Background: Family caregivers of PALS/CIs are highly burdened and at great risk of psychological sequela. Professionals working with these families can be negatively affected on their well-being and are at risk of burnout.

Design: The design was a qualitative interview study.

Methods: One focus group and 10 individual semi-structured interviews were conducted with seven family caregivers and nine professionals after the death of a PALS/CIs. The analysis was guided by the interpretive description methodology and the theory of sense of coherence. This study adheres to the COREQ guidelines and the ICMJE recommendations.

Results: The family caregivers' challenges regarding coping with everyday needs related to the sick person were associated with 'Accepting that nothing else matters', 'Adjusting to new roles while balancing', and 'Realizing different values in relationships'; whereas the professionals' challenges were related to 'Collaboration a balancing act', 'Working in a home of sorrow', and 'Coordinating threads to tie'.

Conclusion: Family caregivers found coping with the complexity of the diseases a challenge, and their everyday life needed constant adjustment to new roles, coping with inappropriate behavior, and navigating through the progression of the diseases of their sick relatives while collaborating with numerous professionals. The professionals struggled with coordinating and collaborating with the families and with other colleagues due to the severeness and complexity of diseases.

Relevance to clinical practice: Findings point to the importance of relationships for caregivers and professionals and a need to provide support through an online palliative rehabilitation program that encompass coping strategies in relation to the diseases.

Trial registration details: Id no. NCT 04638608.

Keywords: amyotrophic lateral sclerosis, caregiver, cognitive impairment, healthcare professional, interpretive description, multidisciplinary, palliative care, qualitative research, rehabilitation, sense of coherence

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Correspondence to:
Lene Klem Olesen
National Rehabilitation
Center for Neuromuscular
Diseases (RCFM),
Kongsvang Allé 23,
8000 Aarhus, Denmark

Department of Public
Health, Faculty of Health,
Aarhus University, Aarhus,
Denmark
leol@rcfm.dk

Karen la Cour
Research Unit of User
Perspectives, Institute of
Public Health, University
of Southern Denmark,
Odense, Denmark

Heidi With
National Rehabilitation
Center for Neuromuscular
Diseases (RCFM), Aarhus,
Denmark

Charlotte Handberg
National Rehabilitation
Center for Neuromuscular
Diseases (RCFM), Aarhus,
Denmark

Department of Public
Health, Faculty of Health,
Aarhus University, Aarhus,
Denmark

Introduction

Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disease that worsens relentlessly, with death occurring after a median of 3.2 years.¹ A review showed that the number of people diagnosed with ALS is increasing, ranging in Europe from 2.1–3.8 per 100,000.² There is now a clear clinicopathological relationship between ALS and frontotemporal dementia.³ Studies show that about 50% of the people with ALS may also suffer from extra-motor signs and symptoms, such as cognitive and/or behavioral impairments, and 10–15% meet the diagnostic criteria for frontotemporal dementia.^{4–6} Family caregivers, like spouses, partners, children, and siblings (hereafter termed caregivers) are described as the pillars of ALS patient care in the literature and research concludes that the consequences of ALS have a profound impact on the psychosocial well-being of these caregivers.^{7–9} Two studies show that behavioral changes are the strongest predictor for psychosocial distress in caregivers of persons with ALS.^{8,10} Caregivers experience decreased well-being due to the more cognitive and/or behavioral impairments than to the physical disability of the person with ALS.^{8,11} Also, health professionals supporting and helping people with ALS are at high risk of experiencing decreased well-being.¹² The presence of executive and behavioral impairments in ALS has also been found to negatively affect the relationship between the ALS patient, family, and the health professionals.¹³ Connelly *et al.*¹⁴ conclude that health professionals need psychosocial support that addresses the moral distress and the ethical challenges related to the terminal neurological illness and the communication performance and focuses on a supportive attitude of acceptance of the inevitability of death.

Background

A review of the cognitive, emotional, and psychological manifestations in ALS found impairments in perception and processing of emotions, social cognition, and alterations in certain cognitive functions, such as executive functions, verbal fluency, language, and verbal memory.³ Moreover, behavioral changes seen in relation to ALS concern disinhibition, apathy, loss of sympathy or empathy, perseverative, stereotyped or ritualistic behavior, hyperorality, or dietary change.¹⁵ Cognitive and behavioral impairments in ALS are associated with more rapid progression and poorer prognosis, with a risk of death being 2 to

2.53 times higher than unimpaired controllers, equal to a median survival of 19–20 months compared to 46 months in unimpaired controllers.¹⁶

Caregivers of people with ALS are often without any previous experience with the role of a caregiver.¹⁷ Caregivers need to accommodate themselves to the significant needs of the person with ALS as well as to a myriad of associated cognitive and behavioral symptoms within a short period of time.¹⁸ Caregivers are known to neglect their own needs and jeopardize their well-being in order to take care of the person with ALS.^{8,9} Rehabilitation assists ALS patients and caregivers to adapt to the psychical and psychological challenges of living with ALS^{19,20} and the palliative care that is essential from the debut of ALS²¹ because it seeks to alleviate psychical, psychological, and existential distress among the ALS patients and the caregivers and improve their quality of life.^{10,22}

Existing research is sparse on which challenges caregivers and health professionals experience in everyday life with the person with ALS particularly in regard to cognitive and/or behavioral impairments (PALS/CIs) and how these can be addressed and supported to enable coping.²³ The aim of this study was to explore reflections of caregivers and health professionals regarding coping with the everyday challenges involved in caring for a PALS/CIs. This effort was undertaken in order to better target future supportive interventions for both caregivers and health professionals.

Methods

Design

The design was qualitative, and the methodology Interpretive Description (ID) was used. ID was chosen as a means to explore the challenges faced by caregivers and health professionals through an inductive analytical methodology.²⁴ ID aims to create knowledge that can help solve challenges that arise in clinical practice.²⁴ The theoretical lens of the sense of coherence (SOC) by Aron Antonovsky was applied to explore the participants' ability to cope with life stressors and how three core components, comprehensibility, manageability, and meaningfulness, need to be present in order to cope with and experience life as coherent and thereby reduce stress.²⁵ Combined ID and SOC guided the whole study from the planning phase, developing of interview guides, and analysis to the writing up of findings by

offering an organizing logic with focus on a practice-based problem, methods to use, and analytical interpretive descriptions within the SOC.^{24,25}

This study was carried out in Denmark, where public healthcare, including assistive devices, is part of the taxpaid National Health System.²⁶ A PALS/CIs can receive help and support at home from either community or private healthcare services without being charged. This study was embedded in the PhD project EMBRACE (FAVN) and affiliated with the National Rehabilitation Center for Neuromuscular Diseases (RCFM).²⁷ RCFM is a highly specialized private outpatient hospital financed by the National Health System in Denmark that supports the public social and healthcare system in rehabilitation of people with different kinds of neuromuscular diseases, including ALS.²⁸ The professionals are organized into multidisciplinary teams consisting of nurses, occupational therapists, physiotherapists, medical doctors, psychologists, and social workers.²⁷ The professionals who work with persons with ALS are called ALS counselor and are provided with the possibility to get access to highly specialized knowledge of ALS and rehabilitation of everyday life with the disease.

Data collection

A total of 16 persons participated: caregivers ($n = 7$) and health professionals ($n = 9$), representing approximately 35 PALS/CIs. The caregivers were two adult children and five partners or spouses. The health professionals were nurses, social and healthcare assistants/helpers, visitation and aid consultants, dementia consultants and occupational therapists, and trained people without a professional education employed in private healthcare services. Hereafter, all are referred to as health professionals. Caregivers were eligible if they were a spouse, partner, or an adult child of a deceased person with ALS and if the caregivers described the deceased as having had cognitive impairments as defined in *International Classification of Diseases 11th Revision*.²⁹ The caregivers were included if they had either been living with the person with ALS until death or had been in a close relationship with the deceased within the last 6 months. Convenience sampling was applied in selecting the caregivers,²⁴ who were asked to contact the first author after an information letter about the purpose of the study was posted on the websites of the patient

organization The Danish Muscle Dystrophy Foundation and the RCFM. The first author then assessed the eligibility of the caregivers according to the inclusion criteria before enrollment. A further two caregivers were recruited through a neurological department at a hospital and one through an ALS counselor at the RCFM (Table 1). Caregivers known to the first author (former ALS counselor at RCFM) or third author (ALS counselor at RCFM) were excluded.

Professionals were eligible to be included if they had a formal experience of caring and supporting PALS/CIs and had either a healthcare education or if they were trained people employed in private healthcare services. Professionals were excluded if they were known by the first or third author. Purposive sampling of the health professionals was applied, aiming for variety among participants in regard to professional background, years of seniority, and experience with PALS/CIs, and their in-depth knowledge by virtue of their equipped experience with the topic investigated.²⁴ Key leaders from specific departments within community health and social care were identified through the municipal administration of one large and one medium-sized community in Denmark, and one leader from a private healthcare service was selected and invited by phone and e-mail. The key leaders chose the health professionals that fulfilled the inclusive criteria (Table 2).

Between March and May 2020, 10 individual semi-structured interviews were carried out by the first author with caregivers and health professionals by in-person meetings in the homes of the caregivers, by phone, or via Skype interviews. Seven caregivers participated in six individual interviews and two related caregivers were interviewed together. Interviews lasted between 57 minutes and 2 hours 7 minutes.

Focus group interview was used with the health professionals in order to allow a dialogue, rethinking, and discovery of unexpected experiences between health professionals from the same community healthcare services in regard to ALS and cognitive impairments.²⁴ One semi-structured focus group interview with five participants and four semi-structured individual interviews were conducted with the health professionals by the first author. The focus group interview took place at local health care center, while the third author observed. Focus group interviews with all the health professionals had been planned but was

Table 1. Demographic characteristics of the caregivers that participated in the study.

Participants – caregivers		(n = 7)
Gender	Male	1
	Female	6
Age	<22	1
	23–45	1
	47–55	2
	56–65	2
	>66	1
Relation	Married/spouse/partner	5
	Adult child	2
Occupational status	working	4
	Retired	3
Years of ALS-trajectory	<1	0
	2–3	6
	4–5	1

ALS, amyotrophic lateral sclerosis.

changed to individual interviews due to the COVID-19 lockdown. The individual interviews with the health professionals lasted between 42 minutes and 1 hour 10 minutes and the focus group interview 2 hours 15 minutes. Ten health professionals were invited, but one nurse dropped out due to extensive workload caused by the COVID-19 pandemic. Two semi-structured interview guides were developed for caregivers (Table 3) and health professionals (Table 4). Interviews were digitally recorded and conducted in Danish, and quotes were subsequently translated into English. The research group consisted of four female researchers: PhD student (master's degree in educational psychology, OT, former ALS counselor), professor (PhD, OT), Senior Researcher (PhD, Master of Public Health, RN) both with experience within the fields of rehabilitation and palliative care, and RN (clinical nurse specialist, family therapist, and ALS counselor at RCFM).

Analysis

The analysis was inspired by the three core elements in SOC: comprehensibility, manageability,

and meaningfulness, which guided an inductive analysis of the participants' way of coping with everyday challenges related to the PALS/CIs.^{25,30} Furthermore, the analysis followed the four-step process guided by the ID methodology.²⁴ First, all interviews were transcribed in verbatim and uploaded into the electronic software program NVivo™ 12. Second, the transcripts were read and initially coded separately by the first and last author, and subsequently the codes were compared according to circumstances found in the data. Third, preliminary relationships were distilled as categories and a critically appraisal of relationship within all data across subgroups and relevant thematic options lead to the primary categorization. In an iterative process of scrutinizing the codes, the whole research team critically discussed the relationships and patterns within the data, and this led to the final extraction of the main messages and capture of the overarching categorical themes. Finally, the last analytic step resulted in the interpretive thematic and conceptual description that formed a model capturing the main findings regarding the caregivers' and health professionals' challenges in their everyday life with the deceased PALS/CIs.²⁴

Ethics

The study was registered on clinicaltrials.gov (Id no. NCT 04638608) and adheres to the COREQ checklist,³¹ the Declaration of Helsinki³² and follows the ICMJE recommendations.³³ The study was not under obligation to notification to the Danish Data Protection Agency (File no. 2019-521-0144), or the Central Denmark Region Committees on Health Research Ethics (File no. 1-10-72-1-19). Participants provided informed consent and both oral and written consent were obtained from all participants prior to data collection.

Results

The analysis with the theoretical lens of SOC gave an overall understanding of the challenges among our participants, the caregivers and health professionals, through six categorical themes.²⁵ The interrelating themes of the caregivers, 'Accepting that Nothing Else Matters', 'Adjusting to New Roles while Balancing', and 'Realizing Different Values in Relationships', revealed how the caregivers were in a constant struggle to understand and capture the meaning and impact of their situations. The themes unfold the

caregivers' difficulties in comprehending and managing the everyday challenges of the PALS/CIs, which caused an overall sense of there being no coherence in everyday life. The relationships with family, friends, and health professionals varied in value and indicated how they either promoted or restrained an SOC among the caregivers. The health professionals acted as an outside interference in the families' everyday lives due to their dependency on help and support, which was represented in the themes: 'Collaboration a Balancing Act', 'Working in a Home of Sorrow', and 'Coordinating Threads to Tie' (Figure 1).

Adjusting to new roles while balancing

The caregivers described how the consequences of their relative falling ill with ALS and cognitive impairments changed the roles in the family and how they had to constantly adjust to these changes.

Yes, but I was still a wife, but at the same time I was also the wife who was doing an ungrateful job . . . I was also the nurse, and all that. Because it was me who was in charge of everything. (ID 8)

As the PALS/CIs became increasingly impaired, the caregivers explained how they had to take greater and greater charge of the everyday chores, taking on new responsibilities in the housekeeping and learning craftsman skills, such as changing tires and fixing things in the home. The caregivers explained how they had to support and help the relative getting dressed, being groomed, guided through daily activities, and moreover help organize and carry out things during everyday life.

I pushed his wheelchair into the bathroom, where his commode was, and then, we went into the bedroom, and I got him undressed and tucked him in. And then in the morning, when we got up, it was all about getting him out of bed and into the wheelchair. And getting him bathed and dressed and getting him downstairs and then we started . . . and I had to give him his medicine and his protein supplement and whatever else he needed in the feeding tube. (ID 8)

They explained how they supported and guided the PALS/CIs during public and social arrangements, where they, in addition, had to deal with provocative outbursts and inappropriate behavior by their partner with ALS, which was caused by the cognitive impairments. One participant

Table 2. Demographic characteristics of the health professionals that participated in the study.

Participants (professionals) in total		(n = 9)
Gender	Male	1
	Female	8
Residence	Urban (>40,000 Citizens)	5
	Rural (<40,000 Citizens)	4
Years of ALS experience	1	3
	2–3	4
	4–5	1
	>6	1
Professional background	*Healthcare professional	8
	Unskilled	1
*Healthcare profession	Nurses	3
	Occupational therapists	2
	Social assistants/helpers	3

ALS, amyotrophic lateral sclerosis.
*Elaborates which professions the healthcare professionals revolves around.

explained how she was confronted by the manager of a grocery shop.

'[Name of the affected partner] is no longer allowed here because he steals'. And I remember how sad I felt . . . And when I came home, I saw . . . [name of the affected partner] had also taken candy from the plastic bins, you know, the ones that are placed at a child's level, and I could also see when he came home, that he was sitting on some candy . . . And I remember being very sad when this happened because there were a lot of people, I knew who were just standing there and staring. (ID 10)

The caregivers spoke about how they had to assume many different roles, informal carer, nurse, project leader, all-round secretary, and caretaker, and at the same time being emotional involved as a spouse, partner, or child.

Yes, she did come to help, and it was partly to let me have time with [the name of the affected partner], but also to be able to have time to do the things that needed to be done besides that . . . Yes, you have many roles in that situation. Yes, but you became a

Table 3. Interview guide for caregivers of deceased persons with amyotrophic lateral sclerosis and cognitive and/or behavioral impairments. (PALS/CIs)

Key questions	Elaborative questions
Try and tell me about yourself	How long has it been since you lost your relative/loved one?
What was your relationship with the person with amyotrophic lateral sclerosis and cognitive and/or behavioral impairments (ALS/CIs) you have lost?	How was your relationship with your family?
Looking back, try to describe what have been most burdensome for you in the whole ALS/CIs disease trajectory?	How did you experience your own situation? What did you do to manage the disease trajectory? What was important for you in this period of life? How did your network react toward you?
How did you become aware of your relative's ALS/CIs?	How did the cognitive impairments (CIs) of your relative affect your relationship? What was your reaction toward the CIs of your relative? How did handle you handle the challenges related to the CIs?
How would you describe everyday life with your relative with ALS/CIs?	How did the ALS/CI trajectory of your relative affect your family? What was your role during the disease trajectory? What kind of losses did you experience during the period? How did you cope with these losses? Who supported you during the trajectory?
Try to explain at which point in the ALS/CIs trajectory of your relative you realized you needed support?	What kind of help and support did you receive? What did you miss in order for you handle the situation? What was the biggest challenge for you in your everyday life? Who was important for you during this period?
ALS, amyotrophic lateral sclerosis; CI, cognitive impairments.	

Table 4. Interview guide for health professionals caring for people with amyotrophic lateral sclerosis and cognitive and/or behavioral impairments (PALS/CIs).

Key questions	Elaborate questions
Try to elaborate on when you felt most challenged in your work with the families of a person with amyotrophic lateral sclerosis and cognitive and/or behavioral impairments (PALS/CIs)?	What was stressful for you in your work with the families? How did you work with the person with CIs? How did you work with the relative to the person with CIs? What would have been supportive for you in this situation? How were you able to take care of yourself? How did you experience your ability to support the carer?
How did you become aware of the cognitive and/or behavioral impairments (CIs) of the person with amyotrophic lateral sclerosis (ALS)?	How would you describe the person with CIs? How did you experience the personality and behavior impairments? How did you experience the language impairments? When did the CIs appear during everyday life? What was your reaction toward the person with CIs? How did the CIs affect your relationship toward the person with ALS? How did you handle the challenges related to the CIs? What was the biggest challenge in regard to CIs? What did you miss in regard to your handling of the situation? What kind of support could have beneficial for you? What was the biggest support for you in this situation? What kind of role did you have in the disease period? When have you felt alone with your challenges?
ALS, amyotrophic lateral sclerosis; CI, cognitive impairments; PALS/CIs, persons with amyotrophic lateral sclerosis and cognitive and/or behavioral impairments.	

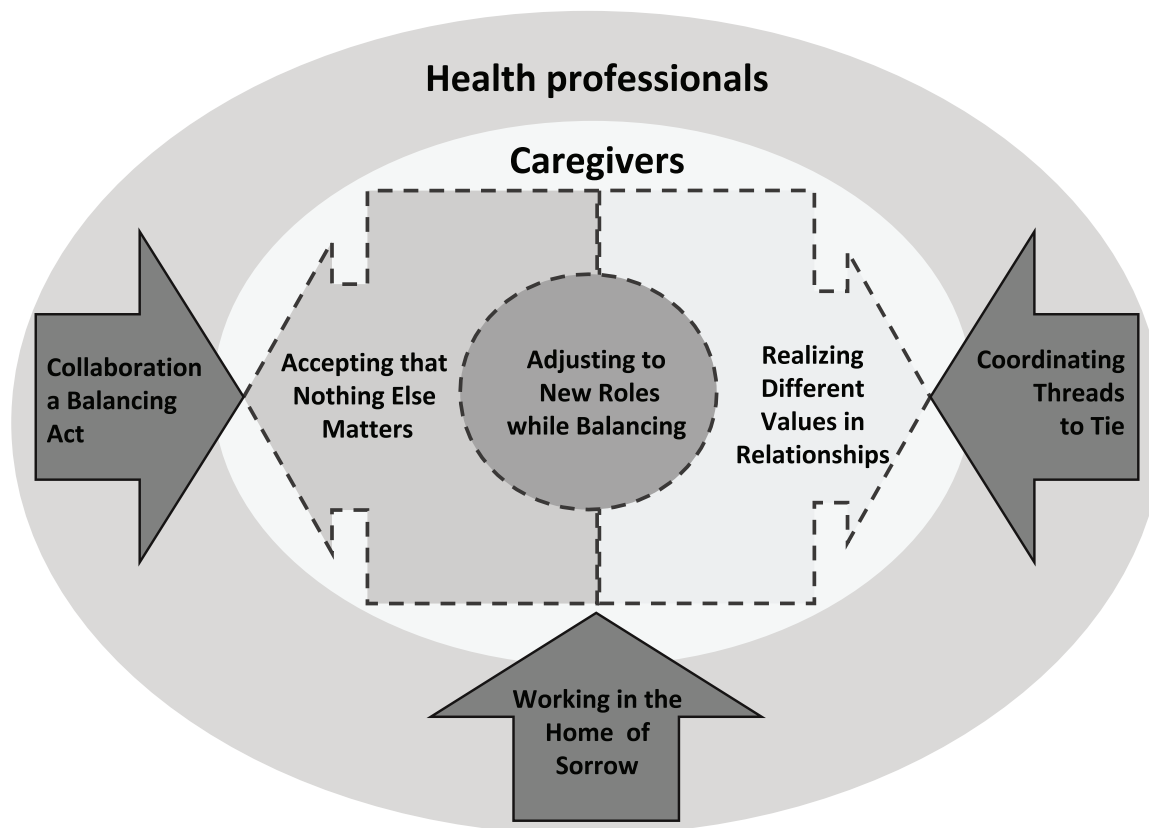


Figure 1. Reflections on everyday challenges of family caregivers and health professionals regarding coping with a person with amyotrophic lateral sclerosis and cognitive and/or behavioral impairments. The findings represent the caregivers illustrated in the inner circle interacting with the health professionals, who act as outsider interference in the everyday lives of the families.

project manager, that's how it was, it became project ALS . . . It did. (ID 13)

The coordinating role in the 'ALS project' was described as a way to get things 'fixed' and as a project that affected every aspect of their daily lives. For instance, they became responsible for the personal as well as the practical care of the PALS/CIs, providing support, coordinating appointments with different departments and people in the hospital and in the community healthcare services, and taking care of the home, kids, economy, and gardening.

The caregivers emphasized how physical impairments affected their relationship and intimacy. They could kiss, caress, and hug and the PALS/CIs only responded passively.

Well, I must say, it does something to the relationship when you have to wipe the saliva off his mouth before you can give him a kiss. No matter whether

you like or not, it does matter! . . . I don't think I really realized it [missing physical contact]. Well, I sometimes gave him a kiss on the cheek, but then I sometimes felt that it was a duty at the end. (ID 13)

Some caregivers expressed how they tried to hold on to the relationship by continuing to sleep next to the person, holding hands, hugging when they helped the PALS/CIs' arm around themselves, but at the same time explained how different and difficult it was.

Actually, right from day 1 when he was diagnosed, it was like that . . ., I thought 'From now on you can't be committed to him as much as you once were a. You may as well start to say goodbye, because he's not staying and whether it lasts 2 years or 5 years or whatever'. I probably didn't expect it to only last [number of] years. It was like that, when he gave me hugs and stuff like that, I did not feel as intensely effected as I once had, and I'm sure, that's because I was starting to let go. (ID 11)

Accepting that nothing else matters

The symptoms from ALS and cognitive impairments overshadowed the caregivers' everyday lives.

I think it's often difficult not to get upset, so I tried all the time to think: 'You're here now and you're trying to make [the affected relative] happy and also make your other relatives happy . . . and then you can wait and get upset when you get back home'. (ID 14)

They argued how time became crucial and limited because they along the course of the development of the disease quickly realized the irreversibility of the condition and that they were about to lose their relative, which emphasized the need to prioritize the time left together. Doing things that mattered to the PALS/CIs, like visiting old haunts, going on weekend trips, and daily excursions became a prioritization of the feeling that nothings else mattered. They described that knowing that it was 'now or never' made them endure their relatives' behavior and sharpen their focus on living in the present moment while putting their own lives on hold.

As my mother said, we were living in the present, it was my father who was top priority, and everyone else had to push everything else aside. It wasn't in his spirit the way things turned out, but that's how it was. And we could only do that because we loved him. (ID 9)

Relinquishing spare time and in general repressing your own needs to support and help the PALS/CIs became a way of coping and making the last period together precious.

Yes, it affected me deeply [when the partner became terminal ill], it did. But I tried as best I could not to show it and then . . . the last 3 weeks [the partner] lived; I almost didn't sleep. (ID 8)

The caregivers expressed that they needed to process feelings like grief, sorrow, and frustration and repress their need to relax, plan their days like before, and sleep properly. The caregivers reflected on their constantly conflicting feelings toward the PALS/CIs. They described how they were divided by their love for their relative, but at the same time, sometimes hoped that it all would come to an end.

I used to say that he died a little every day, and I broke down a little more every day. So, I was just so

. . . , had come to grips with the sorrow before he died, because I'd lost him slowly along the way after all. (ID 11)

They explained how they sometimes wanted to get away from home because their lives were turned upside down and filled with uncertainty, frustration, and sorrow, but as soon as they went to visit friends, they wanted to go back home and not waste the precious time they had left with the relative. The caregivers elucidated how they were torn by conflicting feelings like uncertainty about the future and wanted to get the most out of the little time left rather than being frustrated about not being able to deal with the ALS and the cognitive impairments and take control of their everyday lives. As part of these inconsistent feelings, the caregivers explained that they felt frustrated, scared, angry, and sad when they began to notice the changes in behavior and language and how the lack of knowledge about the disease caused even more conflicts in the families.

I think it was very scary. And I was like 'It's really weird' but it was also when I learned something about it, I think it helped me a lot, but I still feel it was very unpleasant. (ID 14)

Changes in behavior and language became a known and recognizable condition that the caregivers explained they had to endure and, for instance, stopped confronting their relative regarding the cognitive impairments related to uninhibited behavior and language. Everyday life was emphasized by the caregivers as full of losses and sorrow because of the constant progression of the disease. Being a caregiver of PALS/CIs was described as a struggling to manage the challenges and new roles without any prior experience of such a complex situation. This caused a sense of loneliness in a situation characterized by chaos.

Realizing different values in relationships

The caregivers described various relationships as a continuum containing both positive and negative aspects, indicating that some relationships became important regarding being able to cope with everyday life with the PALS/CIs and were therefore considered a lifeline in contrast to adverse relations being burdensome, frustrating, but also sometimes necessary. Relationships with family members, friends, and professionals spanned the whole range of these feelings. The caregivers described how some professionals

became important to the families because they helped and supported them during the trajectory.

We have a care coordinator, [name of the coordinator], and SHE IS SIMPLY WORTH HER WEIGHT IN GOLD. I LOVE HER MORE THAN ANYTHING ELSE IN THE WORLD. And she simply stood by us right from day 1 . . . she took care of everything. Everything that had to be applied for and all that stuff. She got the needed signatures, filled out the papers, made sure to send them, and before I could say, Jack Robinson, she was here with it. (ID 8)

The professionals were the nurse, dementia consultant, general practitioner (GP), neurologist, and health coordinator, who kept calling, asking if they needed any help, prioritized extra time to talk with the caregivers despite having little time. The caregivers described how these health professionals became a lifeline, highlighting the major and vital role the health professionals played regarding the caregivers' ability to deal with changed life conditions. But some had negative experiences, and one caregiver said,

Well, every time we got a new application that had to be submitted, you had to start all over. And you know that as the diseases progresses. You get new social workers who must process the application for speech impairments, for example, or who must take care of the assistive aids or . . . We also had to deal with a new category when he began to need a personal assistant. So, we kept meeting new people . . . And that is also very annoying when you are dealing with a terminal patient. (ID 13)

The caregivers described how the collaboration sometimes became a burden which consisted of hard and sometimes very frustrating work when, for instance, they had to draw the line in relation to how they wanted the professionals to act in their homes, or how they had to fight for the needed help from the community healthcare consultants. The caregivers sometimes felt an increased workload when community or private healthcare services entered the homes because the caregivers had to deal with a large turnover of staff coming into their homes and spend time and energy instructing health professionals, translate the wishes of the PALS/CIs, or rearrange the positions in bed or wheelchair after the health professionals had left the home. They elaborated how this caused insecurity and frustration for

both the caregivers and the sick relative because it was crucial that the health professionals knew the PALS/CIs and how the person should be helped and supported despite their lack of ability to express themselves. The caregivers described how persons in their family and friends played an essential part in relation to their experiences of burden and ability to deal with the disease trajectory.

She [a friend] helped me with many legal things, like applying for disability cards, and, or, you know . . . a disability car, and for compensation for critical illness, and my own critical illness, and . . . And my sister too, and our boys. They've been nothing but amazing, our boys . . . They sometimes kindly said 'Mom, go to a café and we'll take care of Dad!'. (ID 10)

Also, these relationships became a lifeline for the caregivers who often felt lonely despite having people – often health professionals – around them all day. The caregivers also described how other acquaintances surprised them by taking responsibility and action independently and supported them, while others disappeared.

And that's a thing in all of this, you lose friendships . . . So, we knew, that we would now be separating the sheep from the goats again. So, that's the way it is! And you know what . . . I don't have anything left over for people who can't figure out whether to call or come by or something, I frigging don't! (ID 9)

Results of the professionals

Working in a home of sorrow

The professionals explained how the homes were characterized by hopelessness because of the ALS diagnosis and how the cognitive impairments caused sorrow in the family and made the professionals' work more difficult.

What does it mean for a human being to be in this psychological state they're in when they have cognitive impairments? A lot of people find it hard to understand that you can't get back to who you used to be. The fact that you can't just get some medication for it or treatment so that you can get your parent or spouse back. When you have changed so radically that you actually become physically and verbally aggressive or without inhibitions. And you lose your understanding of the disease and try to stand up, and also become scarred of everyone

around you because they don't help you. It's really difficult. These are the biggest challenges I face. (ID 6)

The professionals experienced how caregivers were distressed, anxious, and highly burdened due to their changed life condition as a result of ALS/CIs' condition. The health professionals described how they tried to support family members in coping with this transformation of the PALS/CIs, but at the same time, were confronted with a trajectory of dilemmas and diverse feelings between the couple like hope and hopelessness, in which they constantly had to navigate without erasing hope or causing further sorrow or grief.

Well, I feel most challenged where I can see that the PALS/CIs is simply getting worse and worse and the caregivers in the family won't acknowledge it. You see the grief in the person's eyes due to being so fatally ill, and the spouse doesn't understand that the person has become this ill or refuses to accept it . . . I think that is so terrible to witness. (ID 15)

The professionals explained that they exposed their ears, bodies, and souls to a lot of tears, frustrations, and helplessness from the PALS/CIs and caregivers even though they were often pressured for time or lacked knowledge on how to support the families. Several of the professionals found it frustrating not to be able to ease the caregivers' sorrows, which caused a self-doubt among them. Some professionals explained how they were able to distance oneself from the emotional commitment.

We get so affected by it [ALS/CIs] because it is so hopeless, and the hopelessness . . . it becomes such a heavy burden to carry. We must, although it may sound a little cynical, we have to take two steps back sometimes to see it all in a larger perspective. We cannot involve ourselves as if we were closely related. We need to maintain a professional approach. (ID 7)

Others explained how they often ended up engaging themselves a little extra because they became so affected by the severity of the situation that it made it hard for them to leave their jobs behind them when they were not working. The professionals nonetheless expressed how relieved they were that they were able leave the home compared to the caregivers who were always there.

Collaboration a balancing act

The professionals described the collaboration with the families as a balancing act due to constant potential dilemmas and conflicts lurking in the family and in the collaboration.

There was this woman with dementia-like behavior, and it was very difficult because, well, she had a friend who had the idea that she could just set up a private helper arrangement, but she couldn't, because she couldn't be both the boss and take part in the other arrangements where she could delegate that role, that wouldn't be relevant because the friend couldn't undertake the task to the degree it was necessary . . . and that was actually a great frustration for particularly the relatives. I really don't think the woman [PALS/CIs] had the ability to understand it anymore, but there was a great feeling of dissatisfaction, and the thing about us is that we simply didn't speak the same language, and the relative, she had some expectations about how she just had to put the employees in the system, and then it went completely, well hotchpotch. (ID 7)

They explained how they had to be careful to use the right words and how a wrong choice in actions or attitudes could create an instant negative and tense atmosphere. Disagreements were often related to a lack of balancing expectations between what the professionals could offer and how this could be matched with the family's needs. Working in a private home while adhering to the health and safety at work act and the relevant legislation also sometimes caused an unequal and difficult collaboration with the families.

. . . for us it is a workplace that must be able to function as such, and for them it is a home and must be able to function as such as well, so I sometimes have to compromise, and sometimes they have to be able to compromise. (ID 15)

The collaboration with the families was also affected and dependent on the personal and professional competences and relations to the family, where the professionals had to navigate while delivering a professional service. The professionals talked about revealing or not revealing their personalities and private lives for the families while still struggling with staying professional in relation to the families.

That is something we talk a lot about. When do we open ourselves up a little bit and take something

more personal into it [the relationship], and when do we keep ourselves completely closed up in order to be able to distance ourselves from some of the difficult things that also are there. But it is also important that we sometimes open up a little and allow something personal into it [the relationship], because otherwise the person you face can't relate to you if you are too professional, then you have to open up for some things and bring something personal into them. You just must remember to close up again when you do not need to be open anymore. (ID 6)

The professionals had to reduce frustrations and avert dangerous situations by supporting the caregivers in handling the inappropriate behavior and language of the PALS/CIs.

And it's from experience, the more experience you have, the easier you get into the family, the easier you can 'talk these symptoms down' and talk about what can be done about the symptoms instead of making them worse. Then it becomes something we can work on, and in that way, we can come up with solutions instead of just seeing barriers all the time. And if his language is inappropriate and becomes very offensive 'well, how do you balance it? And how do you respond to these actions? And if you stay, if he's aggressive and lashes out at someone, how do you position yourself by the bed, so he doesn't hit you?' But he can still have his outburst without it hurting anyone. (ID 6)

Various coordinating threads to tie

The professionals referred to a union of different authorities, departments, and professionals that supported the PALS/CIs. They mentioned internal collaborators in the community healthcare settings, like nurses, social and healthcare helpers/assistants, visitation and aid officers, and dementia consultants. Moreover, they mentioned external collaborators like neurological departments, pulmonary departments, GPs, the palliative units, and the private healthcare services. Working harmoniously with the other disciplines was explained by the professionals as both important and useful in regard to fulfilling the complex care needs of the PALS/CIs.

I have experienced, when you have these collaborative meetings where both patient and caregivers are present, and you represent a professional skill, that it can calm down these families, because then they know that everyone has

heard the same thing. And if there is a relative who is so frustrated and would like to split the professionals, then you can say, 'Well, at the meeting where we were all present, so and so and so . . .' (ID 3)

When collaboration succeeded, the professionals described how they supported the families despite different areas of focus, places of location, responsibilities, and lack of acquaintances. For instance, authorities and providers working together to formulate schemes that will secure the ability of the PALS/CI to stay as independent as possible or professionals reaching out to spar with collaborators.

That's where the collaboration works really well with caregivers and with the home care, that's a good support, indeed! Then you experience that, yes, that there is a unified whole and we share common ground, and we can see that we can do this together. We can deliver good support and care for the family, despite all things. (ID 7)

Respect for each other professional expertise, statements, and heterogeneity was also addressed as an important aspect in the interdisciplinary collaboration. The many different professionals helping the families initiated a need to coordinate actions for the professionals to co-operate in and across sectors instead of working in parallel or counteracting each other, which unfortunately was described to be the case in some homes.

Yes, there were several things where I had to make ends meet here and wondered, 'is it me or what is happening?' But the external collaborator simply felt it was a competition regarding who should get to know this PALS/CIs best. Where I finally said to a colleague, 'Well, I do! Because I've been there the most'. Because I'd been there for 4 months before she came, and we have figured it all out together. So, that was MY biggest challenge down there, the respiratory team. (ID 2)

Collaboration became challenging when responsibilities were unclear or when lack of knowledge about or respect for each other's job area was unclear, which affected their job satisfaction negatively. The professionals stated that a coordinator was crucial for both collaborators and the families when trying to cope, plan, and make everyday life as manageable as possible because such person had a close collaboration with and knowledge of how the PALS/CIs and the families functioned.

Discussion

The everyday challenges of the caregivers and health professionals in our study were compounded by the spiraling of losses because the diseases and adaptation were ongoing processes. The challenges initiated inappropriate behavior and constant need for adjustments in roles and support and consequently with a high workload in everyday life. The caregivers in this study were without any previous experience regarding supporting their relative with ALS/CIs. De Wit *et al.*³⁴ found that dealing with behavioral changes without an understanding of symptoms and causes may be most challenging for caregivers because behavioral changes are the strongest predictor of psychological distress in caregivers of people with ALS and are also associated with lower perceived control over caregiving. Research has shown that caregivers of people with ALS have unmet needs regarding support, help, and understanding of their situations.³⁴ This relates well with our findings that showed how the caregivers struggled with adjusting to new roles and realizing the meaning of relations, which could be explained by the lack of comprehensibility of the situation among caregivers.²⁵ In order to cope with the constantly reoccurring challenges during everyday life, the caregivers accepted that nothing else mattered, which seemed to add to their comprehension of coherence and provided meaningfulness and a way to manage and comprehend their complex situation.²⁵ Burke *et al.*³⁵ argue that reconfiguring existing roles within the family could reduce frustration or irritation if cognitive impairments are present. This was also highlighted in a study on the burden of caregivers, where adjustments of roles and responsibilities, interaction style, and activities of daily living may be beneficial and reduce the experience of burden and increase the quality of life for the caregivers.³⁶ A study on the experiences of caregivers of people with ALS regarding care and support concluded that caregivers often stop seeking or accepting support.³⁷ This was also described by the caregivers in this study, but they nevertheless realized the importance of relations with health professionals and their other relatives. Online services could accommodate the need for support among burdened participants, who had difficulties in leaving their relative at home alone due to the extent of caregiving responsibilities. A previous report highlight online services, like telehealth as a way to support caregivers of people with ALS.³⁸ The use of social media have shown to increase the connection between caregivers of people with

ALS, and the attendance and socialization.³⁹ Caregivers expressed trouble with managing cognitive and/or behavioral impairments because they had no knowledge of the disease but nonetheless emphasized how they acquired the needed skills along the way. These findings are in line with a study where caregivers described a reduction in unmet needs as they became more confident and gained more experience as time went on.⁴⁰ This emphasized how the caregivers' experiences improved along the ALS/CIs' trajectory and moreover their comprehensibility that made them capable of managing different tasks and thereby becoming more confident and finding a meaning in the situation.²⁵ De Witt *et al.*⁸ concluded in a study on care demands among caregivers of people with ALS and progressive muscular atrophy that caregivers might feel that they were fully capable of fulfilling care tasks while at the same time experiencing that they have no control over their lives in general. Being emotionally involved as a carer and a novice within such a complex situation has a major negative impact on the well-being of caregivers of PALS/CIs.^{8,35} Due to the complexity and lack of time for these caregivers, telehealth could meet their need for information and support, and Geronimo *et al.*⁴¹ found that caregivers of people with ALS and professionals generally view telehealth as favorable. Targeting educational programs with specific variables to manage behavioral problem is advocated for caregivers of people with dementia.^{42,43} Blended psychosocial support for caregivers of people with ALS also shows a significant effect on self-efficacy related to control over thoughts.⁴⁴

In relation to the health professionals, it was emphasized how working in a home of a PALS/CIs was filled with sorrow and mourning due to constant loss and how it affected their work-life balance because of the hopelessness and the effect on the caregivers. Oxford neurologists, Brian Matthews (1920–2001) likewise recognized a balancing act between the needs of the ALS patient, the carer, and the physician, and he stated that the suitability of specialized neurology practice was about being able to continue to support a case of ALS.⁴⁵ According to the SOC theory, it could be argued that the professionals experienced the sorrow in the family as unmanageable, making it difficult for them to cope with it.²⁵ The professionals in our study explained how they struggled with the mixed emotions, and for some, it was difficult to leave their jobs behind when

they went home. These findings are underlined by the results from a study on professionals working in the palliative field that concluded that professionals are at risk of burnout, moral distress, fatigue, and affected clinical decision-making due to unexamined emotions and unprocessed grief.⁴⁶ Some of the professionals in our study described different ways of addressing the families' need for support, but also highlighted this as a complex and difficult challenge. The professional's explained that their interactions in the homes appeared to matter regarding how the families experienced everyday challenges and their ability to cope with them. These findings are supported by a review on health services that suggests staff training and service delivery models as strategies to remedy problems because skill deficits in professionals can reduce the well-being of the caregivers.⁴⁷ Oliver and Turner¹² likewise found education in the palliative approach useful for professionals caring for a person with ALS and supporting the caregivers. They also stated how a strong multidisciplinary team is important because of each health professional's expertise in the team contributing to a robust internal support to serve the person with ALS and the families more effectively.¹² This is in line with the findings in this study stressing the challenges the health professionals were burdened with in their daily practice – trying to collaborate and coordinate in a home filled with grief and sorrow – a task that needs specially acquired skills and support.

Methodological considerations

The study included 16 participants, which is a relatively small sample size, but the participants had special expertise and knowledge within the research field and the interviews were conducted in depth, which helped broadening the perspectives on challenges in everyday life with a PALS/CIs.⁴⁸ However, due to convenience sampling of the caregivers, results may not necessarily reflect the distinctions in perspectives between caregivers and this could therefore be a limitation.³¹ The caregivers were included based on their personal description of cognitive and/or behavioral impairments of the deceased PALS/CIs, and no disease-specific cognitive screening measure was applied. Having used a specific cognitive and/or behavioral screening measure may have ensured caregivers of PALS/CIs with a higher degree and presence of the cognitive and/or behavioral impairments seen in the ALS spectrum.⁵ However, defining and diagnosing ALS/FTD is widely discussed and

international guidelines from Strong *et al.*⁴⁹ found the term *frontotemporal spectrum disorder* (ALS-FTSD) as most appropriate to characterize the breath and severity of the frontotemporal dysfunction that can be encountered in relation to ALS. Neuropsychological testing is time-consuming, and a variety of simple tools have been developed to assess cognitive and/or behavioral impairments, and some of these are administered by carer.⁵⁰ A review on screening instruments on cognition and behavior in ALS patients in clinical practice showed that not all patients were able to complete a full neuropsychological battery due to motor and/or language impairments,⁵¹ and thus screening in regard to inclusion would be difficult and uncertain. The participants were included based on their specific experience with cognitive and behavioral impairments in the person with ALS who not necessarily had a confirmed clinical assessment or diagnose. Since diagnosing and defining cognitive and/or behavioral impairments in relation to ALS is complex, our results should be seen in this light. In addition, our participants were specifically asked to present challenges related to cognitive and/or behavioral impairments which may reflect a larger amount of these challenges.

Using focus group interviews for the professionals facilitated the rethinking and discovery of unexpected perspectives regarding the health professionals' own challenges in relation to supporting the families.²⁴ The professionals had different experiences of caring for PALS/CIs and different interactions with the families, which caused one especially experienced participant in the focus group interview to be more outspoken, which could have reduced the value of the group discussion and the credibility of the results.⁵² Nevertheless, the interviewer made sure that everyone got the possibility to express an opinion throughout the interview. Having to replace the second planned focus group interviews with the health professionals with individual interviews gave an opportunity to get more in-depth knowledge from participants. Furthermore, only one professional from a private healthcare service was included, and this could be another limitation because these health professionals have different qualifications due not having a health-professional education but being personally trained and often working alone for many hours. However, the participants in our study had many years of experiences with different PALS/CIs and could therefore elaborate on everyday challenges based on a wide perspective.

The results mirror everyday reflections of caregivers and health professionals in a Danish context, which may affect the external credibility owing to a tax-financed healthcare system in Denmark; however, similar results are expected to be found among caregivers and health professionals in other Western countries delivering healthcare services to PALS/CIs. Applying the SOC theory and the ID methodology during all processes in the study helped explore the SOC among participants coping with everyday challenges related to the PALS/CIs in a systematic and stringent way that ensured transparency and internal credibility.^{24,25}

Conclusion

Everyday challenges of caregivers and health professionals of PALS/CIs were found to be affected by the complexity of constant progression of the diseases and the various needs to fulfill. Being emotional involved as a carer and having to struggle with cognitive and/or behavioral impairments without prior experience was found to be the strongest predictor for psychosocial distress among caregivers. The caregivers are heavily burdened by the constant need to adjust to new roles while dealing with unclear responsibilities. Supportive educational interventions, telehealth, and social medias are beneficial among caregivers and health professionals who look after people with ALS because they increase self-efficacy, socialization, accessibility, and reduce burden. The health professionals struggled with mixed emotions and their ability to support and engage adequately with the families without taken home the sorrow from the families. Professional's interaction with families of PALS/CIs had a major influence on the family's ability to cope with everyday challenges and remedy problems. Multidisciplinary approaches and education in palliative care would be useful in creating a robust internal support for these families and at the same time support the health professionals' ability to create a positive work-life balance. This study adds to an understanding of the challenges and needs of caregivers and professionals and the relationship between these. It also highlights elements of support and knowledge that might be important and necessary to incorporate into the development of a targeted online intervention for caregivers of PALS/CIs and professionals who help them. Relevant elements to include in palliative rehabilitation for carers and health professionals could be psychoeducation, peer-support, and knowledge on change in roles, loss of control,

intimacy, preparation for the future, coping with loss and sorrow, and on how to take care of oneself. Future research should address the need to develop and test the outcomes of a targeted online palliative rehabilitation intervention for caregivers and health professionals in order to support their coping with challenges in everyday life with a PALS/CIs.

Relevance for clinical practice

- Caregivers experience imbalance in their relationship due to increasing loss of their partner and challenging needs to fulfill without any previous experience within this role.
- Health professionals struggle to support families while balancing their professional relation toward these and need specialized support and knowledge to fulfill this challenge.
- Caregivers and health professionals are challenged by the trajectory of ALS and cognitive and/or behavioral impairments and are in need of support and knowledge from other health professionals recommended through a targeted online palliative rehabilitation intervention.
- Elements to include in such an intervention could be palliative rehabilitation, psychoeducation, peer-support, and specialist knowledge on changes in roles, loss of control, intimacy, preparation for the future, coping with loss and sorrow, and taking care of oneself.
- Online interventions could increase the accessibility for caregivers and health professionals and thereby meet the gap between the participants' need for support and their lack of time.

What does this paper contribute to the wider global clinical community?

- Knowledge on the need for both a palliative and rehabilitation intervention to help caregivers of people with ALS and cognitive and/or behavioral impairments confront and cope with their challenges.
- Knowledge on important elements to address when developing a targeted online palliative rehabilitation intervention for caregivers and health professionals.
- Knowledge on the importance of strong personal relationships for caregivers and

professionals when being a part of the challenging care and support for a person with ALS and cognitive and/or behavioral impairments.

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Author contributions

Lene Klem Olesen: Conceptualization; Data curation; Formal analysis; Funding acquisition; Investigation; Methodology; Project administration; Resources; Software; Validation; Visualization; Writing – original draft; Writing – review & editing.

Karen la Cour: Conceptualization; Data curation; Formal analysis; Methodology; Project administration; Resources; Supervision; Writing – review & editing.

Heidi With: Conceptualization; Data curation; Formal analysis; Investigation; Project administration; Supervision; Validation; Writing – review & editing.

Charlotte Handberg: Conceptualization; Data curation; Formal analysis; Funding acquisition; Investigation; Methodology; Project administration; Resources; Software; Supervision; Validation; Visualization; Writing – review & editing.

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ORCID iDs

Lene Klem Olesen  <https://orcid.org/0000-0003-0225-6045>

Charlotte Handberg  <https://orcid.org/0000-0002-1378-2449>

Data availability

The data that support the findings of this study are available on request from the corresponding author. The data are not publicly available due to privacy or ethical restrictions.

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