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Experiences of next of kin to patients with amyotrophic lateral sclerosis using invasive ventilation via tracheostomy

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ABSTRACT

Objective: To investigate the experience of being the next of kin to patients with amyotrophic lateral sclerosis who use invasive ventilation *via* tracheostomy.

Methods: Semi-structured interviews with eight next of kin were conducted and analysed using qualitative content analysis.

Results: Three main themes comprising a total of nine subthemes emerged from the analysis: *A turbulent care process aiming to extend life*, *Struggling to cope with the strains of everyday life*, and *Conflicting roles as next of kin and carer*.

Conclusion: The results highlight the importance of involving next of kin throughout the whole care process and considering their specific needs. Furthermore, the development of specific support interventions to facilitate the everyday life for next of kin and to ease their burden are much needed.

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Amyotrophic lateral sclerosis; caregiver experience; invasive ventilation; tracheostomy; qualitative content analysis

► IMPLICATIONS FOR REHABILITATION

- It is important to involve next of kin and consider their needs throughout the whole care process regarding invasive ventilation via tracheostomy.
- Specific support interventions need to be developed to facilitate the everyday life for next of kin and to ease their burden.

Introduction



Being a next of kin and caring for someone with a fatal neurodegenerative disease such as amyotrophic lateral sclerosis (ALS) is often associated with a substantial burden [1,2]. It is reported, from both Sweden [3] and other countries [2,4] that many next of kin to patients with ALS provide a large amount of unpaid care, i.e., are informal caregivers. In Sweden, all citizens may apply for home care services or personal assistance to reduce the impact of disability, regardless of the illness concerned. It is, thus, possible for next of kin to be employed by society and thereby become a formal caregiver, e.g., a personal assistant.

Due to the progressive nature of ALS involving increased muscular weakness, patients will inevitably suffer respiratory impairment at some point. European guidelines on ALS management [5] state that respiratory insufficiency should be managed e.g., through mechanical ventilation, which can be non-invasive (ventilation *via* a mask) or invasive *via* tracheostomy (TIV). Non-invasive ventilation is considered the primary option and, thus, TIV might only be used when non-invasive ventilation is insufficient [5]. It is known that TIV can prolong life by years in patients with ALS [6–9] but also that it may increase the burden of care in next of kin [10,11]. The use of TIV in patients with ALS differs both

between and within countries, e.g., from Italy it is reported to occur in 11% [7] and 31% [6], in Sweden in 8% [12] and in Japan in 33% [9]. A review of the subject found that the rates of TIV ranged from 0% up to 45% depending on the health centre and the country [13]. Clinical guidelines highlight the uncertainties regarding the impact TIV has on patients' quality of life and their caregivers' burden of care [5,14]. Further, the issue of the costs of maintaining the treatment are also raised [5]. The initiation of TIV is reportedly influenced by the attitudes of physicians [15], patients [9,16–18] and their next of kin [16,19]. An important factor, emphasized by both patients and their next of kin, is concern about the effect TIV might have on quality of life [16,19].

The use of TIV in Sweden varies but it is estimated that 8% of the patients with ALS living in the major urban region have TIV [12]. Local guidelines in Stockholm County regarding procedures for initiating TIV state that the neurologist at the ALS clinic should refer the patient to the Respiratory Centre for surgery if clinical, cognitive and social conditions are fulfilled [20]. The Respiratory Centre handles the aftercare and educates patients, their next of kin and formal caregivers, such as personal assistants, as to how to manage the ventilator, the tracheostomy tube and the stoma.

There is a lack of studies on experiences of next of kin to patients with ALS using TIV. Most studies investigate the

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experience and life situation of the next of kin to patients with ALS using non-invasive ventilation [21–23]. To the best of our knowledge only three qualitative studies exist on the experiences of next of kin to patients with ALS who use TIV [11,24,25]. These report that the next of kin's quality of life decreases, that caring is a dynamic process, and that it is important for next of kin to find the prolonged life of the patient meaningful [11,24,25]. The results from these few studies might not be transferable due to differences in health care contexts. For example, Swedish next of kin may be employed and become paid personal assistants which might influence their experience. Knowledge of this kind of experience is needed if health professionals and social services are to anticipate and address important factors concerning the home-based care of patients with TIV. Thus, the aim of the present study was to investigate the experience of being the next of kin to patients with ALS who use TIV.

Materials and methods

Participants

Participating next of kin were recruited *via* patients with ALS who used TIV and were registered at an ALS clinic in an urban region of Sweden. A total of 12 patients were identified in May 2017 and all, but one, according to their neurologist were regarded as suitable to contact. Thus, 11 patients received mail with information about the study and were asked to identify one or more next of kin who could communicate in Swedish and could be approached by the researchers. A total of eight next of kin were identified of whom three were related to the same patient. Next of kin were informed of the study both orally and in writing. Informed consent was obtained at the start of the interviews prior to data collection. Table 1 presents the characteristics of next of kin ($n=8$) and patients with ALS ($n=6$), and Table 2 presents information on participants providing quotes. The study was approved by the Regional Ethical Review Board, registration number 2016/1086-31 and procedures were conducted in accordance with the Helsinki Declaration.

Table 1. Characteristics of next of kin ($n=8$) and patients with ALS ($n=6$).

Next of kin	
Age, years, median, (IQR), min-max	49 (35–58), 27–64
Sex, women/men, n	6/2
Relation	
Wife, n	3
Mother, n	1
Father, n	1
Daughter	2
Son	1
Cohabiting with patient, n	4
Work status	
Employed as personal assistant full time/part time, n	3/4
Patient with ALS	
Age, years, median, (IQR), min-max	55 (31–62), 26–68
Sex, women/men, n	1/5
Time since diagnosis, years, median (IQR), min-max	5.0 (4–11) 3–12
Invasive ventilation since, years, median (IQR), (min-max)	3.5 (1–6) 1–7

Data collection

Data was collected by face-to-face interviews at a location chosen by the next of kin, most often in their home. Each interview was conducted by two data collectors (CY, KG, or PS), of whom one led the interview and the other could follow-up on important matters. A semi-structured interview guide was developed and started with the question “Can you tell us about an ordinary day in your everyday life?” This main question was followed by additional questions aiming to go deeply into the experience of the next of kin to a patient with ALS and TIV, e.g., “How have things changed since TIV?”, “What was the situation like coming home from the hospital?”. All the interviews were audio taped and transcribed verbatim.

Analysis

To analyse the experiences of next of kin to patients with ALS and TIV, as well as the underlying meaning of those experiences, a qualitative content analysis with an inductive approach [26,27] was carried out. Manifest and latent content analysis was performed, according to the steps presented in Table 3. Authors CY and KG analysed four interviews each (step 1–4, Table 3), continuously discussing the results. Authors PS and MK took part in the analysis of one interview each (step 1–4, Table 3) and discussed the results with authors CY and KG. The analysis consisted of comparisons between sections and the whole of the interviews with a process of repetition back and forth. To address the question of trustworthiness, the process of analysis for steps five and six was conducted by authors CY and KG together. The themes and the subthemes were discussed and refined at meetings with all the other authors. The interviews were anonymised during transcription and analysis thus, confidentiality was maintained.

Table 3. The steps of the content analysis process.

Step 1.	The transcribed interviews were read through several times to get a sense of their content.
Step 2.	The text was divided into meaning units comprising words, sentences or paragraphs related to each other by their content and context.
Step 3.	The meaning units were condensed, i.e., the text was shortened without interpretation and still contained the core of the meaning units.
Step 4.	Through a process of interpreting the underlying meaning, each condensed meaning unit was labelled with a code.
Step 5.	Codes were compared and those with similar content were grouped into subthemes. The initial number of subthemes was reduced after a process of re-reading codes and re-considering the interpretation of the underlying meaning and the whole context, thus moving between the whole and parts of the text.
Step 6.	The subthemes were analysed by their latent content in a process of investigating how next of kin experienced invasive ventilation via tracheostomy in their relative with ALS. Three main themes were constructed based on the underlying meaning of the subthemes.

Table 2. Information on participants providing quotes.

Next of kin 1 > mean age and cohabiting; patient > median time since diagnosis and TIV
Next of kin 2 < mean age and cohabiting; patient > median time since diagnosis and TIV
Next of kin 3 > mean age and cohabiting; patient < median time since diagnosis and TIV
Next of kin 4 < mean age and not cohabiting; patient < median time since diagnosis and TIV
Next of kin 5 < mean age and not cohabiting; patient < median time since diagnosis and TIV
Next of kin 6 > mean age and cohabiting; patient > median time since diagnosis and TIV
Next of kin 7 > mean age and not cohabiting; patient < median time since diagnosis and TIV
Next of kin 8 < mean age and not cohabiting; patient < median time since diagnosis and TIV

Results

Three main themes emerged from the analysis, comprising a total of nine subthemes. The first theme was **A turbulent care process aiming to extend life** and its subthemes were: "Feeling insecure in a tense emergency situation", and "A roller-coaster care process". The second theme; **Struggling to cope with the strains of everyday life**, had the following four subthemes: "Putting one's own life on hold", "The challenge of coping with life-giving TIV", "Reliance on personal assistance needed to feel secure", and "The struggle to communicate when the power of speech is lost". The third theme was **Conflicting roles as next of kin and carer** and its subthemes were: "Being an informal care-coordinator", "Contradictory relationship roles", and "A desire for support and guidance in the carer role". The main themes with subthemes, illustrated by the participants' own words, are described below.

A turbulent care process aiming to extend life

Feeling insecure in a tense emergency situation

Next of kin described how TIV as an alternative had often been discussed before the situation became acute, and in some cases the intervention had been planned. Despite this, the surgery was nonetheless carried out as an emergency in connection with a quick decline in the ability to breathe. That the decision was finally made in an emergency situation was experienced both as relief at avoiding a long-drawn-out process, but also as a forced-decision where next of kin felt a lack of sufficient knowledge. That the doctors repeated the question about initiating TIV on several occasions in the emergency situation was experienced as important, but also as emotionally exhausting. In the light of the uncertain prognosis when TIV was initiated, whether the patient would survive surgery, and how the near future would be, the next of kin experienced the emergency situation as chaotic and frightening. The situation was described as scary and full of forbidden thoughts about life and death. They felt that it was impossible to discuss the alternative namely not to choose TIV.

No, somehow it was not allowed, he must live. One shouldn't think about whether he would die. (Next of kin 1)

Next of kin were unsure as to whether the patient chose TIV based on a desire to keep on living without being influenced by them. Next of kin insisted that the decision regarding TIV was the patient's own, and that they, as being close, supported whatever decision was made.

You mean if I was for or against it? After all, I was involved when we made that decision so ... it was about his decision. If he didn't want it, I would have supported that. Now he wanted it, so we support his decision. It is his life and his decision. (Next of kin 5)

Regardless of information about the difficult situation that TIV would result in, for the next of kin there was no other alternative. They described how TIV meant that the patient could carry on living, but also how it meant understanding impending death.

When one undergoes this operation, it is like saying that I accept that I cannot breathe, this is how I shall die, of this illness. So, it feels a little ambivalent namely that this is a life preserving treatment that gives better quality to the life that one has left, but that one would rather be without. (Next of kin 8)

A roller-coaster care process

The care process after TIV was initiated was perceived as a roller-coaster journey, i.e., there were ups and downs with feelings of relief, uncertainty and fear. It was a relief for next of kin when the

patient was taken care of in the intensive care unit. This enabled them to let go of responsibility for the patient's health.

I thought that it was comforting that he was admitted where they really could keep an eye on him and knew what was to be done. So, although he was in intensive care I was at ease, it felt good. Then we knew that, well OK, they'll take care of him now. Then one could relax and know that there's not much more to be done for us, they're doing their best here. (Next of kin 4)

The period after intensive care in different hospital departments was, however, described as of low standard with inadequate support and information, and uncertainty as to future care. It was a turbulent period and as a consequence of shortage of beds, the patient was often moved around between different departments. The next of kin remarked that the staff's competence and attitude towards the patient's prognosis varied considerably between different departments. Sometimes they felt afraid to leave the hospital and that they were responsible for compensating for the staff's lack of skill.

We simply didn't dare leave because she was not in good hands, no it was the most frightening thing I have experienced. I don't understand how it could be that way in hospital, in a specialist unit. (Next of kin 7)

It was said to be a relief when the patient finally came to the Respiratory Centre where both skill and treatment methods were felt to be professional. The period of hospitalization after TIV was initiated and before a return home was possible, was described as surprisingly long. It involved extensive preparations in the form of training and recruitment of personal assistants, ordering of supplies, and running through of routines.

I thought that it was hard that he remained in hospital for quite a time. This was since all the staff were forced to go through training in order for him to be able to go home; so that dealing with the ventilator could be delegated to the personal assistants. It meant that he had to remain in hospital for two months when he wasn't really ill anymore, except for his Motor Neuron Disease, but he had to stay. (Next of kin 8)

Returning home was experienced by some next of kin as safe, while others described a difficult process with a shortage of personal assistants.

Struggling to cope with the strains of everyday life

Putting one's own life on hold

Next of kin pointed out that it is impossible to imagine what it is like to have someone with TIV at home, and to understand how immense the necessary adjustments are. They described how their own life had been put on hold, and that how their situation was at the limits of what they could cope with. Their own daily life had changed and been adapted to the patient's needs.

It is as if my life was at a standstill because of the way the situation is. One works, eats, sleeps; that is evidently all one has time for. (Next of kin 5)

Next of kin described a changed daily life in which the patient's comfort was always their focus. Moreover, this was at the forefront of their thoughts, regardless of how involved they were in the daily care. They were constantly on hand and prepared to intervene in an emergency.

You never know when something might happen, so it is a question of being available all the time and you jump whenever the phone rings. (Next of kin 4)

Next of kin reported being constantly attentive to the patient's comfort and constantly aware of the noise from the ventilator.

You have to keep track of the breathing all the time, you hear the alarm from the ventilator and we have a sound monitor too, if the tube slips out or something like that. (Next of kin 3)

Being available throughout the day and night had a negative effect on both the sleep and stamina of next of kin. They described their situation as feeling as if they were constantly using a reserve battery, and that there was no margin or space for personal interests or activities. Their home had been changed into a hospital and the sense of home comfort had been badly affected by the noise from the apparatus and machines. The constant presence of personal assistants imposed limitations on private life making it difficult to relax as well as resulting in limited opportunities to withdraw.

You can't have a peaceful moment at home, there are staff there all the time, care going on all the time. (Next of kin 2)

At the same time, some next of kin described how daily life carried on with new routines. They had got used to this changed daily life in which their own activities had to be planned for or taken away entirely.

The challenge of coping with life-giving TIV

In the view of the next of kin, TIV was the decisive intervention making the patient's continued life possible, and they felt much gratitude for that. Their perspective had shifted from a belief in the patient's imminent death to thinking that the end was not so close.

None of us believed that he would live as long as he has in fact. Before I believe that we thought more that by Christmas he'll be dead or that by the summer he'll be dead. But now, I don't think at all any more. (Next of kin 6)

Some regarded TIV as one aid amongst others and felt that the ALS-disease itself affected life more than TIV. Others saw the life-giving TIV as terrifying and were constantly afraid that something would happen. Nevertheless, insofar as the patient's face was more visible with TIV (in comparison with the breathing mask) care was also made easier.

The next of kin felt that they had better strategies for dealing with the patient's breathing problems following the specific education and training associated with TIV which involved distinct routines. They described the importance of this structured care and that TIV demanded great carefulness and attentiveness in the light of everything that could happen.

One is terrified that the tube will get stuck in something, one really is, that the canula will slip out. There have been situations where I have had to act very quickly; cough, use the balloon, shift the mucus and then loosen it up and suck it out. You are scared evidently but it is a question of acting fast and correctly just then, it just has to be done. (Next of kin 3)

It was imperative for a sense of security in this situation that next of kin knew how to respond correctly when the TIV alarm went, and that they had gained experience with respiratory care. However, several of them reported that they carried out care in the way they thought was best for the patient, and not necessarily according to instructions, as for example in the suctioning of mucus. Depending on the patient's different needs, respiratory care was integrated with all the other kinds of attention, but entailed additional strain due to such things as low-quality disposable materials and incorrect deliveries.

Reliance on personal assistance needed to feel secure

A well-functioning situation with regard to the personal assistants in the home was a prerequisite for the next of kin to be able to

feel secure. It was difficult to recruit enough personnel to provide for the patient's need for round-the-clock care. Recruiting personal assistants, who had the competence needed to care for patients with TIV, was also a challenge.

Much is expected of the assistants; we have to have assistants who can cope with this. There were people, who were excellent assistants, but who couldn't quite cope with the language and couldn't benefit from the training; they couldn't manage when everything became more advanced. (Next of kin 4)

Difficulties in recruitment of personnel could result in the next of kin having to work irregular hours as a personal assistant and being expected to fill in at short notice – in the case of sick leave for instance. The next of kin pointed out that the personal assistants needed to be reliable and careful. According to next of kin, this meant that they had to understand how important it was to manage the TIV equipment correctly and to follow the care routines, as well as being sensitive to the patient's needs and wishes. In addition to being able to satisfy needs arising out of respiratory care, the personal assistants were required to be able to respond to all the other different needs the patient had. This included being able to communicate with the patient in a satisfactory way and to take part in recreational pursuits.

The next of kin thought that there was a lack of sufficient knowledge and competence among the personal assistants despite the special training they had received for TIV. Thus, the next of kin felt a constant need to be on the lookout to be able to compensate for this inadequate care. The question of which of the personal assistants came in the morning was crucial insofar as a reliable person made it possible for the next of kin to take a break from listening to the ventilator in order to spot problems in time. Interacting with the personal assistants in the home and getting them to perform their tasks in a satisfactory way without creating conflicts could be experienced as difficult.

I do try, I think to myself, say it nicely and in a friendly way, don't forget to sign or look now how much more saliva has come, think how much more there has been of late. One has to try not to make them feel guilty, that they have done something wrong, but it is very hard. (Next of kin 1)

It happened that personal assistants had to be replaced since some of them could not live up to next of kin's demands for safety measures in the management of TIV.

With some, just now one boy, one feels: no, we would never trust him. He doesn't have the right feel for the job. You have to understand the seriousness. Meticulousness. (Next of kin 3)

The next of kin highlighted the importance of an effective team leader so that their personal assistants functioned as a team and that they could rely on one another. Some next of kin felt that some personal assistants were too involved and came too close, whereas others regarded it as self-evident that the personal assistants should take part in all the social activities in the home. In some cases, the personal assistants were thought to be fantastic and the next of kin felt safe in handing over responsibility to them.

The struggle to communicate when the power of speech is lost

Next of kin described how communication became one of the most difficult things when the patient's ability to speak had gone, and how this involved a feeling that the patient had changed as a person.

What has been absolutely the hardest with the whole of this thing is that he has lost the power of speech, that is the difficulty in communicating. It is really tough. Not to be able to walk is one thing, but not being able to communicate... (Next of kin 8)

Communication *via* the computer or with yes/no questions took a long time and meant impoverished communication lacking in emotion and gave rise to uncertainties as to whether next of kin understood the patient correctly.

He can write sms and mail, and then there is something that you have to understand, that you cannot ... that is what he writes is very brief and sort of literal. So, you can't put so much feeling into what they say, these sms. (Next of kin 8)

Next of kin described the different technology aids as essential for every-day communication with the patient, but they also felt a certain resistance to beginning to use these changed means of communication.

We got a new eye computer a month ago, but it is difficult. We need to be better at trying to use it. And we haven't been champions at that. (Next of kin 3)

The eye computer was said to be an important aid which made it possible for the patient to ask for help and made leisure pursuits such as surfing on the Internet and social interaction *via* social media possible. It became harder to use, however, when the ability to move the eye declined, which was frightening and meant increased uncertainty in communication. Whether or not the technology worked was crucial and sometimes problems arose because of limited computer capacity. Next of kin wanted the personal assistants to be familiar with, and to use the communication aids, but they also stressed the need for sensitivity and skill in reading and interpreting non-verbal communication.

Conflicting roles as next of kin and carer

Being an informal care-coordinator

Being next of kin to a patient with TIV meant a heavy work-load with many parallel tasks such as responsibility for the patient's care side by side with acting as a spokesman for the patient in time-consuming contacts with the hospital and the authorities. The next of kin wished for more proactive contributions from the health-care system so that the initiative did not have to come from the personal assistants or next of kin. They described how things would have been easier if someone else had co-ordinated the care of patients with TIV.

Co-ordination with the medical care that would have been so wonderful, if one only had had a person who could be sort of the spider in the web, a co-ordinator. Constantly having to call, keeping telephone times and getting hold of people, and then having to explain and so on. If only there had been a person who could help with that, or even the care professionals themselves, so that it had been more co-ordinated. (Next of kin 4)

One area for improvement wanted was a better and quicker process for estimating the need for advice about when to use various technical aids such as eye computers. Some next of kin were very pleased with the access they had to new aids when the patient's condition deteriorated quickly, while others reported that the aids arrived when it was too late for them to be used.

See that you get aids before you need them. In our case it has been too late the whole time. The eye computer after he was able to use it any more, PEG when he couldn't swallow, the electric wheel chair when he couldn't walk any more, instead of being able to use these things more as aids than as musts. (Next of kin 3)

Another suggested area of improvement was the digitalisation of communication between the medical carers, the authorities and patients with ALS when, for instance, the patient could no longer open an envelope or write his/her signature.

There was considerable variation in how contacts with the health-care system and the social services were experienced, with regard to availability and the way next of kin were treated for example. Next of kin expressed a wish to be listened to and a desire to avoid conflict when discussing the care of patients with staff at primary care centres and in-home care.

Less prestige would be good then, we have had massive problems, they want things just their way and that doesn't work here, of course we are very accommodating, but when they come and altogether try to charge over us then it doesn't work. (Next of kin 5)

Some next of kin were very pleased with the primary health-care staff's commitment, while others were dissatisfied with a lack of flexibility regarding times for home visits and a lack of staff continuity.

The next of kin felt that safe respiratory care was important, and they were satisfied with the regular contacts with the Respiratory Centre where the necessary competence and skills were to be found. They were pleased with the education in TIV offered at the Respiratory Centre, but would have welcomed getting respiratory related training earlier, before TIV started. They experienced the specialist neurological clinics as being focused mostly on neurological investigation and the ALS-diagnosis along with carrying out yearly checks and providing certificates.

Contradictory relationship roles

Next of kin wanted to be able to keep their relationship as i.e., wife or son and wanted to hold back from the care of the patient. This was made more difficult by the fact that they had to work a lot as personal assistants and to cover for absence.

I sort of want to come here and be next of kin, come here when one chooses to and not because – now you must. (Next of kin 5)

Working as a personal assistant and being a close relative at the same time was described as hard-going with unclear boundaries between work and leisure. It was difficult to separate the roles as next of kin and personal assistant, and it was also difficult to let go of responsibility. The next of kin reported that their role was to represent security and to keep up a light-hearted atmosphere in the home, as well as helping the interaction with the personal assistants. When the patient's basic needs had been met by competent and reliable personal assistants, there was more space for offering emotional and social support.

I wish in some way that I was not so involved in it, that I could take a step back. When the assistants have been good, when it works, then I feels that it is more this, yes, how shall I say, feelings. Not the usual exchange but more how is he feeling, try to make it so he feels as well as possible. (Next of kin 1)

Most next of kin described how their relationship with the patient had changed for the worse and amongst other things had meant that the patient's dependency on them had increased. But, the serious situation with TIV and the challenging nature of communication had in some cases also meant that they had become closer and that they felt a need to sort out problems and the misunderstandings that occurred quickly.

You get closer to each other in a completely different way. At the same time, for me as healthy, this was not really what I had expected. (Next of kin 6)

Relations with other family members had been negatively affected; the pressing situation in which the patient was the focus of all attention and concern sometimes meant that other family members were neglected.

What has been absolutely toughest is the family relationships in this, because it is an enormous strain for everyone. (Next of kin 4)

The next of kin emphasised the importance of communication between the members of the family as well as respect for one another's life and opinions. The circle outside the closest family had narrowed, friends and next of kin had partially stopped making contact and there were limited possibilities for socialising and travel.

A desire for support and guidance in the carer role

Next of kin felt that their own life situation was very tough, and that they needed more specific psychological and social support in relation to TIV. Some next of kin experienced the psychological support they received as inferior while others answered that they could have had support if they had wished and had had the strength for it. They were of the opinion that an offer of contact with a professional with special knowledge of the circumstances arising from ALS and TIV should be routine.

That there was some kind of a social worker, who came to us all the same and asked, Ok but how are you feeling, without our needing to be almost forced: now it is time for a meeting because we need to look over this, it's part of the routine. Not so that we must actively say no to anyone, but being forced a little - in a nice way. (Next of kin 4)

There was a need to talk about how hard it was to accept ALS as a fatal illness and to reveal taboo thoughts regarding what life would have been like if TIV had not been chosen. The future and the possible scenario of stopping TIV was a source of anxiety for the next of kin. They wondered whether one can decide to stop TIV and in that case how that is gone about.

When and how do they do it? What does one do? Yes, I don't know. It's terribly difficult. Do they pull out the cord to that apparatus then, then he suffocates. Or what does happen, I have absolutely no idea. (Next of kin 8)

Next of kin were worried about the consequences of the patient's reduced or lost power of communication in relation to possibly stopping TIV. At the same time as they wanted to avoid such frightening and inhuman thoughts about the future; they felt the need for the support of a neutral person in preparing themselves and planning to end TIV.

Next of kin expressed a desire to get help with prioritizing what was important in daily life. In addition, they wanted support and advice about how to deal with fear and ignorance as well as how, as carers, they were to respond to the patient's need for aids. They also needed concrete guidance regarding social aspects such as housing and living conditions.

If he does not want to live at home, if he is very sick or something, what possibilities are there then? The hospital will accept a few months or days, but not that one lives there. That's why I wonder about future possibilities. I just want to know. (Next of kin 2)

Next of kin wanted support in thinking through their circumstances and their right to sometimes put their own interests before those of the patient. To cope with the situation, they used such strategies as going out for a while, creating a space for their own social activities and going away if that was possible.

If I feel that I can't cope I go out and get some fresh air. If I am at home they call all the time, can you help. It's the whole time. (Next of kin 2)

Discussion

The findings of this qualitative study provide an important contribution to the limited knowledge on the experience of next of kin

to patients with ALS using TIV. It is unique from a health care context where next of kin may be employed and become paid personal assistants. Three themes emerged from the interviews describing the experiences of next of kin in terms of *A turbulent care process aiming to extend life; Struggling to cope with the strains of everyday life; and Conflicting roles as next of kin and carer.*

Although TIV had been discussed, most next of kin reported that tracheostomy was performed in an emergency situation. This is, however, not unusual, as acute tracheostomy is reported in up to 67% of those using TIV [7,15,28–30]. The next of kin stated that the decision regarding TIV was the patient's own and that they fully supported this decision. Nevertheless, to be involved in the decision and at the same time stand on the side lines in the tense emergency situation was described as emotionally demanding. In line with previous reports, next of kin described the decision making regarding TIV as resembling a choice between life and death that induced existential distress [16,19]. It has been shown that interventions such as specific advanced palliative care planning [31] and structured education programmes on mechanical ventilation [32] can facilitate discussions between patients with ALS and their next of kin. Moreover, the decision-making process is furthered by a reduction in anxiety [31,32]. Discussions on prolonging life and end-of-life issues should be initiated sensitively and at suitable times [31]. It has been recognised that for an empathetic situation to be achieved health-care professionals may need increased communication skills for these discussions with patients and next of kin [31,33]. Thus, in discussions with patients and their next of kin it is crucial to consider the existential concerns raised by the initiation of TIV.

There are regional guidelines for a planned care process for TIV, including initiation and follow-up [20]. However, since tracheostomy was performed in an emergency, these guidelines could not be followed. This might have contributed to the perceived roller-coaster journey with feelings of relief, uncertainty, and fear described by next of kin. Thus, there is a need to develop a prepared and structured care plan also when TIV is initiated in an emergency situation. Next of kin should be involved and the pedagogical aspects of information exchange need to be considered [34] to ascertain that they understand information regarding the care processes in relation to TIV.

The present study shows that next of kin struggled to cope with everyday life and that their own life had been put on hold. All their focus was on the patient and his/her well-being. These results coincide with previous studies reporting that next of kin experience a substantial physical and emotional burden and a low quality of life [10,11,25,35–38]. The next of kin in the present study emphasized the extensive disruption of the home situation as TIV intruded into the family's life. The magnitude of impact on the living situation, such as major changes in the home environment with medical devices and personal assistants working there, might not have been clear to them. Further, having TIV in the home environment induced feelings of "being on duty" 24-h around. For the next of kin to be better prepared, individually adapted information about inevitable changes in the home situation should be supplied as well as regular follow-ups making it possible for them to reflect upon, and if needed, acquire tools to cope with or adjust to the new situation. Research is needed on how this information and support can be organised and delivered.

Next of kin described how they were very grateful for TIV which made the patient's continued life possible; so that the end was not so close. In line with previous studies next of kin described how valuable it was to be trained in preparation for

home ventilation [11] and how they had wished for respiratory-related education even before TIV [32]. Although next of kin had acquired better strategies and felt more secure in dealing with the patient's daily breathing problems, some regarded life-giving TIV as terrifying while others regarded it as one aid amongst others. Before TIV is initiated, it is therefore important to communicate to next of kin how TIV can be experienced differently.

The regional guidelines state that patients with ALS using TIV in home environment require two personal assistants to provide for the patient's need for round-the-clock care [20]. However, personal assistants in Sweden do not need any formal health-care education, which could be considered a requirement. This meant that next of kin experienced specific difficulties such as recruiting competent and reliable personal assistants, being attentive to inadequate care, and having to fill in at short notice. For next of kin to feel more safe, personal assistants need adequate competence and skills to handle the complex needs of patients with ALS undergoing TIV.

Next of kin described how impoverished communication, when the patient's ability to speak had gone, involved e.g., struggling to cope with technical communication aids and being fearfully uncertain as to understanding the patient. Since communication difficulties were described as one of the hardest things, facilitating the initiation and continuous use of communication aids, such as an eye computer, with proper timing should be a main priority.

Having to coordinate the care and acting as a spokesman in time-consuming contacts with the hospital and the authorities was described by next of kin as a heavy work load. The majority of the next of kin in the present study were employed as personal assistants and they described an unclear line between formal and informal care. They expressed difficulties in keeping their private roles, such as being a spouse, a parent or a child, when they also were caregivers. Since the patients' basic care needs had to be prioritized in the demanding care situation, there was seldom space for being "merely" a next of kin. Although being a next of kin and caring for someone with ALS can entail satisfaction and positive experiences [3,11,39,40] it often is a substantial burden [1,2,10,11,25,35,38] and may strain the relationships. It has previously been reported that next of kin feel obliged become informal caregivers [11]. Thus, next of kin to patients with ALS undergoing TIV may need support in making more informed decisions about caregiving to reduce their strain. The next of kin highlighted a need for specific support from a professional with insight into what it means to be next of kin to patients with ALS and TIV. Psychosocial support should therefore be offered, including the possibility to discuss issues ranging from caregiver roles and housing, to existential issues such as what happens at the end of the patient's life. The results from the present study may inform the development of interventions to support next of kin.

A uniqueness of this study is that all eligible patients with TIV were asked to invite a next of kin to participate. This resulted in variation regarding sex, age and relationship to the patient amongst the participating next of kin. However, the fact that three next of kin came from the same family and thus, had a larger impact on the results, might be seen as a limitation. Although the sample size might be considered small, participants were recruited from the region with almost one fifth of the total Swedish population and the interviews resulted in rich data and was considered sufficient for the analysis. That interviews were conducted by two data collectors, of whom one could follow-up on important matters, is a strength and possibly gave richer data. Similarly, the trustworthiness of the results increased from the

frequent discussions between the authors when analysing the codes into subthemes and themes.

In conclusion, being next of kin to patients with ALS using TIV involved experiences of a turbulent care process in the acute phase, struggling to cope with the strains of everyday life, and of conflicting roles as next of kin and carer. The results from the present study highlight the importance of involving next of kin throughout the whole care process and considering their specific needs. Furthermore, the results can be used in the development of specific support interventions to facilitate everyday life for next of kin and ease their burden.

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