

## RESEARCH ARTICLE

# The experiences of patients with amyotrophic lateral sclerosis of their decision-making processes to invasive home mechanical ventilation—A qualitative study

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## Abstract

**Aim:** To explore and gain knowledge of the experiences and needs among patients with amyotrophic lateral sclerosis (ALS) of their decision-making processes whether to choose invasive home mechanical ventilation or not.

**Design:** A qualitative study.

**Methods:** A phenomenological-hermeneutic approach influenced by Ricoeur's interpretation theory was used. Seven patients with ALS were interviewed. The Consolidated Criteria for Reporting Qualitative Research checklist was used for reporting.

**Results:** Three themes were evident in patients' accounts of the decision-making process: (1) being taken care of directly after receiving the diagnosis, (2) living in uncertainty about what the future would bring and (3) doubt causing patients with ALS to change their minds. Patients with ALS were burdened with everyday life challenging decision-making processes about future treatment and doubt caused patients to change their minds about their future treatment. It is necessary to support patients in their decision-making processes using shared decision-making.

**Patient or Public Contribution:** No Patient or Public Contribution.

## KEYWORDS

amyotrophic lateral sclerosis, decision-making process, invasive home mechanical ventilation, mechanical ventilation, non-invasive home mechanical ventilation, patient experience, phenomenology and hermeneutics, qualitative studies, shared decision-making

## 1 | INTRODUCTION

Amyotrophic lateral sclerosis (ALS) is a disease being rapidly progressive affecting patients' whole-body function and has an impact on them psychologically. Therefore, patients at an early disease stage have to be well informed about their future treatment opportunities:

home mechanical ventilation (HMV) which is either non-invasive or invasive or palliative care. It is important that they are able to make an informed decision before it becomes too late and they end up with invasive home mechanical ventilation (IHMV) without wanting it. Therefore, it is important to uncover aspects that may ease the patients' decision-making process.

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## 1.1 | Background

The disease, ALS, is a rapidly progressive neurodegenerative disease that predominantly involves upper and lower motor neurons (Morris, 2015). It is a rare disease with an incidence in developed countries of about 1–7 per 100,000 per year (Thomas et al., 2018). In Denmark, the incidence is 1–3/100,000 and the prevalence is 3–7/100,000 (Sundhed, 2020). The mean age of onset is 58–63 years of age (Andersen et al., 2018). The disease is characterized by muscular weakness and atrophy, and patients' functioning is affected significantly over time, for example when talking, swallowing and breathing (Valko & Ciesla, 2019). Over time, patients with ALS are in a need of support for their everyday routines. The disease is progressive, currently without a cure and there is no effective treatment to halt or reverse the disease. Generally, suffering from ALS leads to death from respiratory failure in 3 years of symptom onset (Couratier et al., 2016). However, survival may be extended if the patient receives respiratory support. In the late stages of the disease, patients with ALS enter a minimal communication state or a totally locked-in state in which the patient is unable to express his or her intentions by any means. This provides an ethical challenge for decision-making about IHMV treatment (Magelssen et al., 2018). In addition to physical symptoms also psychological changes (hope/hopeless, depressive symptoms etc.) were important and it is estimated that around 50% of patients suffering from ALS will develop frontotemporal dysfunction or dementia during the course of the disease which is characterized by neuropsychological deficits with cognitive and behavioural changes such as apathy and disinhibition (Ranganathan et al., 2020). These changes might also influence patients' decision-making process.

It is important that patients are well informed about their options about future treatment. This includes making difficult decisions about the quality of life, dignity and survival (Greenaway et al., 2015). In one study of patients' experiences of living with ALS, patients experienced their lives as a 'count down' where they were in need of support from their family to reconstruct their lives (Yuan et al., 2021). Further, a scoping review has highlighted concepts addressing the complex needs of patients with long-term tracheostomy with or without mechanical ventilation. They made it clear that further research is needed about healthcare quality and outcomes (Stark & Ewers, 2020).

Living with the incurable disease, ALS, is a huge challenge for both patients and their relatives, as relatives can also experience being burdened due to the disease influencing their home and the everyday life of the patient (Winther et al., 2020). Patients with ALS and their relatives have to make several decisions during the trajectory of the disease, one of which has to do with mechanical ventilation.

In a review from 2017, it was shown that the decision-making process about IHMV treatment is very complex. Due to mental and physical dilemmas, it is important to give adequate information to patients with ALS (Danel-Brunaud et al., 2017). In a review of qualitative studies from 2019, different experiences of decision-making

about IHMV were found (Ortenblad et al., 2019). Ortenblad et al. (2019) found that some patients with ALS felt coerced to accept treatment with IHMV, while in other studies, life with IHMV was described as being dominated by worries and insecurities. Furthermore, difficulties were experienced in relation to adjusting to necessary and comprehensive monitoring, involving a 24-h assistance team, and this gave rise to a sense of diminished autonomy and continued concerns (Ortenblad et al., 2019). A study has shown a decrease in quality of life when patients received unwanted treatment with IHMV (Vianello et al., 2011). Studies have also shown that patients with ALS often end up receiving unwanted treatment like IHMV, and the main reason for that is that patients deliberately postpone the difficult decision-making process until it is too late (Murray & Butow, 2016).

So far, studies have described the burdens of living with ALS and IHMV. Further, studies have outlined ethical challenges when receiving treatment with IHMV. To our knowledge, no studies have examined the everyday lives of patients suffering from ALS when deciding whether to choose IHMV in a Danish setting.

## 1.2 | Aim

The aim of this study was to explore and gain knowledge of the experiences and needs of patients with amyotrophic lateral sclerosis in their decision-making processes whether to choose invasive home mechanical ventilation or not.

## 2 | METHODS

### 2.1 | Study design

This study used a phenomenological-hermeneutic approach in order to derive patients' experiences and perspectives when deciding about whether to choose IHMV. The study design was influenced by the philosopher Paul Ricoeur's theory of interpretation to gain a deeper understanding of the experiences and perspectives of the lives of patients with ALS and at the same time elicit new knowledge about being in the world when living with ALS. Ricoeur has argued that phenomenology and hermeneutics in combination can provide a deeper understanding (Ricoeur, 1976). Semi-structured interviews influenced by Kvale and Brinkmann (2015) were used to elicit the findings. The Consolidated Criteria for Reporting Qualitative Research (COREQ) were used to ensure accurate and complete reporting (Tong et al., 2007).

### 2.2 | The Danish setting

In Europe, there is wide variation in the provision of IHMV (Dybwik et al., 2010) and in relation to delivering care and treatment and to whom, depending on the healthcare system in the country.

Most of the patients suffering from ALS in Denmark live at home with either non-invasive H MV or IH MV (Dreyer et al., 2014). The Danish setting differs from other countries with respect to two aspects: assisted respiratory surveillance and daily care. With the Danish publicly financed health system, there is statutory support for formal caregivers of patients with ALS, both with respect to their assisted respiratory surveillance and their daily care. A Danish study showed that survival from disease onset increased for patients treated with IH MV. The average survival rate increased from 22.9 months with no treatment to 56.8 months in patients treated with non-invasive H MV followed by IH MV (Dreyer et al., 2014). Usually, discontinuation of treatment is determined by the patients themselves and is often associated with patients being close to 'a locked in state' (Dreyer et al., 2014). In this study, we will explore patients' experiences and needs when deciding about whether to choose IH MV or not and aim to understand their challenges during the process.

### 2.3 | Setting and participant selection

The study was carried out at Respiratory Centre West at Aarhus University Hospital, which is one of three respiratory centres in Denmark. Recruitment took place in collaboration with a project nurse and a clinical nurse from the centre who identified and selected the patients.

To obtain variation in the experience, the selection was based on purposeful sampling with respect to time after diagnosis (in years), current treatment and the chosen treatment for the future, either with or without IH MV (Polit & Beck, 2018). Inclusion criteria were men and women, all ages, diagnosed with ALS, a period of time (from months to years) after ALS diagnosis, who needed to decide between non-invasive H MV or IH MV, patients who were in contact with the centre and Danish-speaking patients. Exclusion criteria were: men and women who already had IH MV and patients with cognitive impairment. The number of participants was estimated to be a maximum of 10, as this was a qualitative study and given the low number of patients in Denmark with ALS. Ten patients were identified as potential participants, three of whom refused to participate as they said that they could not manage it, and due to having a busy life due to various examinations at different hospitals. Seven patients agreed to participate. The participants were contacted by telephone or at the department. If a patient showed interest in participating in the study, the patient was contacted by the first author, and verbal and written information was given.

### 2.4 | Data collection

Semi-structured interviews were used to obtain descriptive information about the phenomenon of interest: patients' experiences, thoughts and feelings in relation to their decision-making processes.

The interview guide included mainly open-ended questions, which were supported by follow-up questions, if needed, and questions from the participants (Kvale & Brinkmann, 2015).

The interviews were conducted at the hospital or in the participants' homes, according to the preference of the participants. Only the first author and the participant were present at the interviews, although at some interviews, a relative was also present. The first author who was previously a nurse in the centre with patient-related care experiences conducted all of the interviews. The first author had not provided care for the participants included in the study. The participants were interviewed from April to August 2018.

The interviews varied in length from 30 to 90 min. The duration of the individual interviews depended on the participants' ability to speak, their need for pauses and how the participants were affected emotionally. A summary was made at the end of each interview for the patient to confirm. All interviews were recorded and transcribed. Transcribed interviews were not, in general, sent to participants for comments, but one participant had difficulty speaking, and the transcription was sent to the participant for validation. The participant added a few points on missing words and details like verbs and sentences describing the experiences of e.g. uncertainty and a changed mind.

### 2.5 | Data analysis

The interviews were coded by the first author using the software programme NVivo version 10 which systematizes data analysis (QSR International Pty Ltd, 2015). The method of analysis used was developed by Dreyer and Pedersen with inspiration from the interpretation theory of Paul Ricoeur. It includes three levels of analysis: a naive reading, a structured analysis and a critical interpretation and discussion (Dreyer & Pedersen, 2009; Ricoeur, 1976).

The naive reading is the first level where the transcribed interviews were read as a whole to acquire a general, first impression of the text, grasping the first meaning and understanding of the text. From the naive reading, a first impression and insight into the experiences of being a person suffering from ALS and having to decide on IH MV or not was gained. The next level is the structural analysis, where an in-depth analysis of the text supported a profound understanding of the text. In this level, the textual material was analysed at three levels: (1) what was said (quotations), (2) what the text spoke about (meaning units) and (3) the emergence of themes. At this stage, analysis and interpretation formed a dialectical process, with movements back and forth between parts and whole, and between understanding and explanation (Dreyer & Pedersen, 2009; Ricoeur, 1976).

See Figure 1 as an example of the three levels in the structural analysis. In the critical interpretation and discussion, the findings were further interpreted and discussed in relation to other studies, philosophy and theory. The research team discussed the findings.

What was said (quotations)	What the text spoke about (meaning units)	Emergence of themes
<i>Yes, the decision so far is... not IHMV. However, I always hurry to say "so far"... Yes, that it is the decision, so far. I am thinking of the day, where I will be a vegetable and just sit here and not be able to do anything at all. I think it will be hard for me. Nevertheless, that is how I think about it now (P2).</i>	Doubt is evident in the phrase "so far", but the patient is still convinced not to choose IHMV, because being like a vegetable and not being able to do anything in the future is not desirable. That is the decision and the reflections about it now. However, it might change over time.	Doubt can cause patients with ALS to change their mind.

FIGURE 1 An example of structural analysis.

TABLE 1 Characteristics of the participants.

Participants	Time since diagnosis (years)	Current treatment	The decision made and stage of the decision-making process	Place of interview	Comments on the interview
P1	1 year	Non-invasive HMV	Decided on IHMV but has doubts	Home	Affected speech
P2	1 year	Non-invasive HMV	Decided on IHMV but has doubts	Hospital	Affected speech. Relative participated in the interview
P3	1 year	Non-invasive HMV (just started)	No decision, but thinks it may be IHMV, though has doubts	Hospital	Relative participated in the interview
P4	6 months	Non-invasive HMV	Decided on non-invasive HMV. Does not want IHMV.	Home	Affected by pathological crying
P5	1 year	Non-invasive HMV	Decided on IHMV, but has doubts	Home	Changed decisions again and again
P6	2 years	Non-invasive HMV	Decided on IHMV	Hospital	Awaiting surgery to have IHMV
P7	2.5 years	Non-invasive HMV	Decided on IHMV, but has doubts	Home	Relative participated in the interview

Note: Time since diagnosis (years): Estimate in years since the participants received the diagnosis until the interview in 2018.

Abbreviations: IHMV, invasive home medicinal ventilation; Non-invasive HMV, non-invasive home medicinal ventilation.

## 2.6 | Ethics

The study followed the applicable ethical principles (World Medical Association, 2013). Anonymity was ensured, and recordings and transcriptions were stored confidentially. All participants agreed to participate and gave oral and written consent. Participation was voluntary, and the patients could withdraw from the study at any time. The study was reported to the Danish Data Protection Agency (journal no. 1-16-02-30-17).

## 3 | RESULTS

Seven patients participated in this study. The characteristics of the included participants are shown in Table 1 below. The findings from the structural analysis revealed three themes: (1) being taken care of

directly after receiving the diagnosis, (2) living in uncertainty about what the future would bring and (3) doubt causing patients with ALS to change their minds. Each theme will be presented with quotes in italics.

The findings of the naive reading are captured in a short story (Table 2). The story captures experiences from all the seven interviews. In presenting first impressions, we try to enhance the reader's understanding.

### 3.1 | Being taken care of throughout the first period of time after receiving the diagnosis

At the beginning of the disease and for some time after, the participants found they had to make many decisions. There was always something to decide:

TABLE 2 Narrative.

The patients experienced the disease as harsh and unfair. The patients were at different stages in their disease trajectory and thus at different stages in the management of the disease. Most patients found it difficult to decide whether to choose IHMV. The patients needed support in the period after the diagnosis was confirmed and the patients doubted their choice and needed better care. Patients found it particularly difficult due to worries and concerns about what the future would bring

*In our situation, we have to decide about everything, do everything on our own, just do everything, at the same time as having all this to do with the disease, and we have to keep this on our mind and think about what is associated with this disease, plus everything else. [...] At the start, I had a hard time with all of this (P7).*

The quote above shows how hard it was for the patient to make decisions about a lot of things while suffering from ALS, especially at the beginning after receiving the diagnosis. This was supported and experienced by another participant: *'The days in the beginning. I remember them clearly... One decision after another. Just thinking about choosing... You know choose between life and death (P1)'.*

In the aftermath of being diagnosed with ALS, one participant wished: *'It is here [in the beginning] that healthcare professionals really need to support us... (P5)'.* Some participants called for more help from healthcare professionals so that they were taken care of during the period after receiving the diagnosis. Another participant mentioned that a doctor told the participant at the beginning of the disease:

*Now you have to go home and then you have to, not tomorrow or the day after tomorrow, but one day you have to work out, and preferably soon, in relation to the time you no longer can breathe. [...] But how was I supposed to be able to take a decision about that at that time? It was just after I received the diagnosis? (P2)*

As the participant mentioned, *'How was I supposed to be able to take a decision'* about non-invasive HMV or IHMV just after the participant received the diagnosis. The following quotation supports this point further:

*We felt when we got home from the hospital with the three letters, ALS and... not much else. It would have been nice, if there was someone who could have taken care of us for a few days, or whatever. [...] Well, someone who could explain it to us or help us with some of those unresolved questions that we had back then. If we had not done something ourselves, it would have been a long time after I was diagnosed until we visited the hospital. [...]. It took a couple of months and that's a long time to be thinking about those things yourself (P2).*

The participant and the relative talked about the need for support and help after they went home from the hospital. They needed someone *'who could have taken care of us'* at the very beginning of the disease – as mentioned in the quote *'someone who could explain it to us or help us with some of those unresolved questions'*. They got help by themselves

because if they did not, there would have been a long period from receiving the diagnosis until the next follow-up at the hospital.

### 3.2 | Living in uncertainty about what the future will bring

The feelings and thoughts that the participants had in relation to living in uncertainty about the future took up a lot of mental space. Amidst all of the uncertainty, the participants tried to find hope in their situation:

*Yes, but it's hard. I cannot look that far ahead at the moment. Because I do not know what's going on? I do not know how things will be when I have IHMV? [...] We have to hope for the best, and yes, the future is unknown... I do not know if I will be here in a year or two, you know, or if I will receive IHMV? We know nothing. But we can hope (P3).*

The word *'hope'* was mentioned often by the participants and their relatives in relation to the unknown in the future. Further, the same participant added the following: *'I can do nothing else. Hope is perhaps my only way through this (P3)'.*

With no knowledge of what the future might bring, is it clear that they tried to find hope in this new situation. This was also clear from another participant: *'Now I have to believe in hope. I can't do anything else (P1)'.* One participant described not being able to do anything about the progression of the disease: *'I'm suffering from a disease that determines everything. [...] I have nothing to say, I just have to see what happens (P2)'.* Another participant experienced a kind of powerlessness: *'I feel powerless with this disease (P1)'.*

The future was uncertain for the participants, so the participants expressed they had to live their lives in the best possible way in their new situation. Uncertainty was mentioned in relation to the future and the participants did not have much time left to live. Uncertainty about when their life would end was described by one of the participants in the following: *'I'm going to die soon... I had expected to be living life now (P3)'* and elaborated on as follows:

*I do not know. So... We do not know how much time I have left, and it is not even certain that we will use the caravan. We know nothing about the future... We just knew that we would retire together, enjoy each other and life... and then... yes, then this disease comes and takes it all from us (P3).*

One participant talked about the future and death as follows:

*One thing is for sure, and it's that I'm going to die. Maybe the day after tomorrow or maybe I have a year? What I do not know is when. The only thing I'm sure about is that I'm going to die... and probably soon (P2).*

The future was uncertain for the participants and the only thing they knew for sure was that death would occur in a few years. Another



participant talked about the future and not knowing about growing old with their relatives. Life would go on for their families after the participants were no longer there: 'My family will live on without me and I... I just want to die in my sleep and have peace (P4)'. Knowing that they will die because of the disease, but not knowing when death would occur and uncertainty about the temporal development of the disease led to insecurity, which is reflected in the quotation below:

*If I was not sick with ALS, I knew that I had a future to live my life and I would be working for a little more and I would go on early retirement. I could start to relax and enjoy life after a life with full-time work and busy days. [...] Of course, I could get sick, yes, I could have got cancer... With cancer, I might have had a different kind of hope and hope to be able to get well, but with ALS... I know nothing about my life with ALS and my future (P6).*

The future in relation to retiring and enjoying life with the family were distant prospects for the participants. After a long life with a full-time job and busy days, they had been looking forward to retiring, but now their plans had changed since being diagnosed with ALS. Another disease may have left the participants with hope as mentioned in the quotation above which refers to cancer, because with cancer they may have had a chance of being cured or getting better. This was further supported by: 'If I was just sick with a lifestyle disease, then I would have taken everything [...]. That way, I had something I could relate to (P2)'.

The relatives hoped for the best for the participant and believed that they would be able to live for years. The participants did not believe and hope in the same way as their relatives. This is expressed in the following: 'With her, I can find hope, in this madness, but it soon disappears again (P5)'. It showed that the relatives found faith and hope in the situation, which benefitted the participant but hope may quickly disappear again. The participant elaborated further:

*We are all different from each other, and some have completely acknowledged that they have me just for a while and, hmm, yes ... My wife believes that I can live quite a few more years. [...] It's really good that she believes in it. After all, someone has to believe in it. [...] But we can be pleasantly surprised, right? For it is hope and belief that it's useful (P5).*

Everyone is different and some people hoped and wished for more than others, like the wife who believed that the participant would live for years. The words 'it is the hope and belief that it's useful' showed how people suffering from ALS found support.

### 3.3 | Doubt causing patients with ALS to change their mind

Participants' experiences of their decisions about future treatment concerning non-invasive HMV or IHMV showed that doubts caused them to change their mind repeatedly:

*Yes, the decision so far is... not IHMV. However, I always hurry to say "so far"... Yes, that it is the decision, so far. I am thinking of the day, where I will be a vegetable and just sit here and not be able to do anything at all. I think it will be hard for me. Nevertheless, that is how I think about it now (P2).*

'So far' was a recurring term for the participants, when describing the decision-making process of their future treatment. The participant above described the decision as being for now, followed by a comment on how the future might be for the participant as 'a vegetable', not able to do anything, just sitting there. The participant was not able to face the decision. This was elaborated on the following: 'I've not finished wondering about it, but the decision right now is as it is (P2)'. It showed that the participants were thinking about the decision to have IHMV, but were not sure about it. One participant was convinced about the decision to get IHMV, but still had doubts about the future decision: 'I will choose IHMV. When the time comes. It can prolong my life by many years. [...] It's just about getting the best out of it. [...] But I do not know (P3)'. Even if the participant was convinced about choosing IHMV, as it was to prolong the participant's life, there was still doubt about the decision, as the participant's statement ended with 'but I do not know'. Another participant said 'Yes, so I said yes to the hole in the throat, you know, a tracheostomy. [...] I have brought it up a bit whether I should or should not (P5)'. From the quotes above, most of the participants included comments like 'So far', 'I'm not finish wondering about it', 'I do not know it' and 'I have brought it up a bit whether I should or should not'. The statements from the participants indicate their doubts about their decision-making processes of the future treatment.

Even if the participants choose IHMV, the disease would still progress, as mentioned by this patient:

*Even if you get a tracheostomy, there are so many other things that break down in your body, and I will not be able to live with that. [...] Finally, it comes to this conclusion that it is not worthy for me, right. Because there is still the possibility, once you have got it [IHMV], to change your mind, but no. [...] Most of us choose to stay alive. But it can also be too hard (P5).*

This participant above decided to have IHMV, but still concluded that it was not so valuable, and that the patient still had the opportunity to deselect IHMV, as patients with ALS always have the option to withdraw their decision.

Two patients had already decided on their future treatment and were clear about their decision. One of the participants decided not to have IHMV:

*Yes, I do not want IHMV and I do not want to be resuscitated and it has been worked out.... It is 100% clear for me. This has been the case all the time, no matter what, I do not want IHMV. [...] IHMV is no cure, I will still have my disease. I'm also thinking of my family... The idea that they have to live on without me... it is... it is hard and it did give me reason to doubt for a while (P4).*

The choice not to have IHMV was clear for one participant, as 'IHMV is no cure'. The participant mentioned that there was a reason to doubt when thinking of the family. The participant supported this with: 'For me, the most important thing is to leave in a proper way... without IHMV (P4)' and further 'We are now in the situation where it is really bad. [...] We do not have a choice at all (P4)'.

On the other hand, one participant mentioned:

*But... I have always said when I think it's worth being here, I want IHMV, and when it's not fun being here anymore, we will turn the IHMV off. So, it's simple for me. Nobody... It was not a decision I made from one day to the next. It was actually something that I considered a lot. [...] Also.... Yes, then I wanted to be here a little longer (P6).*

The participants, no matter if they decided to have IHMV or not, all experienced doubts during their decision-making processes and experienced changing their minds about the decision.

### 3.4 | Methodological considerations, strengths and limitations

The selection of the participants should be mentioned as a limitation as it turned out that three patients did not want to participate in the study because they said that they lacked the energy. This may indicate a selection bias of participants with the greatest energy. Furthermore, relatives were present at some of the interviews which might have caused some participants not to reveal all of their experiences to avoid hurting their relatives. Our study has drawn data from a small study population. We interviewed a group of ALS patients at different stages of their disease after diagnosis (in years), but all were in the process of deciding whether to choose IHMV. So, although it was a small study population, we gained important and varied information from the participants. There was variation in time after diagnosis in years, current treatment, decision-making about the type of mechanical ventilation (non-invasive HMV or IHMV), place of interview, information about the impact of the disease on their body and whether a relative participated in the interview. We would therefore argue that we gained information-rich data relevant to the problem under study (Polit & Beck, 2018). From an international perspective transferability of the results might be limited due to the differences in cultural understandings to withdraw of life-prolonging treatment (Magelssen et al., 2018). In some countries, it is for example not considered ethical to remove a ventilator once it has been fitted, and discussions based on the withdrawal premise are no longer possible.

## 4 | DISCUSSION

In our study, we found that the very beginning of the disease trajectory might have been difficult time as many decisions had to be taken by the patients and their relatives. The patients expressed a

desire to be taken care of especially throughout the first period after receiving the diagnosis. A scoping review found that patients with ALS and other motor neuron diseases need empathetic and supportive attitudes from healthcare professionals (Oh et al., 2015). Another scoping review also supports this, identifying a significant need for more practical, social, informational, psychological, physical, emotional and spiritual support (Oh & Kim, 2017). We found that several participants requested being cared for in the first period because they found it difficult to relate to decision-making right after receiving the diagnosis.

According to Elwyn et al. (2017), it may be useful for healthcare professionals to focus on the three-talk model, which is a model for learning how to achieve shared decision-making. The three-talk model may be necessary to focus on in the future for healthcare professionals, as the statement above says something about timing. The three-talk model prepares the patient for talking about choices (Elwyn et al., 2017). According to our findings, it is also about timing; it should not be too early in the disease trajectory or too late where the patient would not have the physical capacity to participate verbally in the decision-making process. A Swedish study showed that when the diagnosis was confirmed, patients and their relatives experienced the sense of losing solid ground, and even though they tried to understand their situation, it was difficult (Ozanne & Graneheim, 2018). Another study has highlighted an association between depression and quality of life in ALS patients shortly after the diagnosis (Jakobsson Larsson et al., 2017). Therefore, it is important that healthcare professionals pay attention to the needs of patients with ALS just after they receive the diagnosis, as the participants in our study described the start as difficult. A review examined care for patients with long-term tracheotomies, with or without mechanical ventilation, and identified key concepts reflecting the existing knowledge of preconditions and facilitators to needs-based healthcare for this vulnerable and complex patient group (Stark & Ewers, 2020). Those findings related to our findings of the importance of 'being taken care of directly after receiving the diagnosis', and the review also identified the importance of multi-professional teams with nurse coordinators, where those coordinators should be involved according to the patients' complex needs and further coordination in collaboration with other health professions (Stark & Ewers, 2020). A study from Canada supports the importance of delivering messages to patients with life-changing diagnoses, such as ALS; in that context, the patients wanted a follow-up interview to be set up and agreed upon with their doctors (Mirza et al., 2019). As our participants were calling for more help from the health professionals throughout the first period after receiving the diagnosis. A Swedish study has highlighted the need to focus on what is most important for the individual in the trajectory of the disease, emphasizing ways of supporting patients with ALS in sense-making (Ozanne et al., 2013).

The participants' feelings and thoughts in relation to living in uncertainty about the future preoccupy them. The participants tried to find hope in their situation. A Swedish study (Ozanne et al., 2013) has examined how patients with ALS create meaning in life, even

though they experience anxiety about the issues of life and death. The patients found meaning in life through their family and friends who supported them and made it easier for them to accept their current situation (Ozanne et al., 2013). In our study, the word 'hope' was often mentioned by the participants and their relatives in relation to uncertainty in the future. Hope might be a way for participants and their relatives to live with ALS. Hope from relatives might support the patients with ALS, which was also found in another study. This study also highlighted awareness of the relatives struggling to find meaning in life (Ozanne et al., 2015). Relatives of patients suffering from ALS experienced their lives to be limited and isolated, where they lived an 'imprisoned life'. The relatives felt lonely and that life was unfair. In addition, many were in mourning over the inevitable loss of their relative in the future, and the study concluded that support should be provided to the patient and their relative (Ozanne et al., 2015). This was also shown in a Danish study where the everyday life experiences of close relatives of patients with ALS were examined (Winther et al., 2020). It indicated that healthcare professionals may offer an all-around perspective in relation to patients and their relatives living in uncertainty about the future. Furthermore, our findings are similar to another Swedish study, which showed that living with ALS leads to uncertainty, with patients and their relatives fearing the unknown, especially just after diagnosis. They both live with fear, constantly trying to find a reason why they had been bothered by the disease (Ozanne & Graneheim, 2018). In our study, we found that due to the lack of knowledge about what the future might bring, patients and their relatives try to find hope in their new situation, experiencing the disease as determining their situation. Also, a kind of powerlessness was found, where the only thing they know for sure is that death will occur in a few years. Consistent with our study, a Cochrane review has shown that, in general, patients using patients' decision aids experience greater satisfaction with their choice of treatment (Stacey et al., 2017). This is due to increased knowledge about treatment options and risks. The Cochrane review also showed that using patient decision aids decreases patients' anxiety and worries about the decision. Another review among patients with serious illness supports these findings. Patients with serious illness benefit from shared decision-making and patient decision aids (Austin et al., 2015).

In our study, we found that doubt caused patients with ALS to change their minds about future treatment with non-invasive HMV or IHMV. Doubt was an integral part of the decision-making process, which is a similar finding to a review that examined the cognitive, emotional and psychological manifestations of patients suffering from ALS (Benbrika et al., 2019), where patients' cognitive, emotional and psychological states changed day by day. Some of the participants in this study wanted to choose IHMV but had not yet made a final decision. They doubted whether the choice they had made was right. Especially when they began to feel that their body was slowly changing, doubts returned. Some participants explained that they would not recover by choosing IHMV, which is why they decided not to have IHMV. Although some patients were clear about choosing IHMV, they still experienced doubts. A Swedish study

showed that family, friends and physical health were important for the overall quality of life of patients with ALS in relation to the development of the disease and reduced body function (Jakobsson Larsson et al., 2017). Jakobsson Larsson et al. (2017) also showed that more patients experienced good quality of life despite impaired physical functions. Their study showed that patients with ALS were able to find the quality of life despite having an incurable disease and reduced physical function, which was also the reason why they chose IHMV (Jakobsson Larsson et al., 2017), and may be one reason for the increasing uptake of IHMV in patients with ALS. In relation to the above, a Danish study showed that in the period from 1998 to 2012, there was an increase in treatment with non-invasive HMV and IHMV (Dreyer et al., 2014). There was also an increase in the period from 2012 to 2016, as stated in a report on IHMV in patients' homes (DEFACTUM, 2017). Most of the participants in our study were thinking about IHMV because they knew they always were able to withdraw their decision or treatment with IHMV at any time. This was also found in another Danish study where healthcare professionals experienced an increase in patients with ALS withdrawing treatment with IHMV (Dreyer et al., 2012). All the participants, whatever their decision about IHMV, experienced doubts and were still experiencing doubt and changing their minds about the decision. It can therefore be discussed if patients with ALS generally lack information about treatment with IHMV and are not adequately supported in their decision-making processes. A Cochrane review has shown that using patients' decision aids reduces doubts or decisional conflicts (Stacey et al., 2017). Our study showed that the patients lacked information or support during the decision-making process, indicating that a patient decision aid might have been helpful.

## 5 | CONCLUSIONS

In conclusion, our study showed that patients living with ALS mostly experienced doubt during the decision-making process about their future treatment. The beginning of the disease trajectory was perceived as unmanageable, with feelings of powerlessness due to living in uncertainty about what the future would bring. Therefore, to be taken care of in the period directly after the diagnosis was important. Lastly, using patient decision aids might improve the shared decision-making process when deciding between non-invasive HMV and/or IHMV.

## 6 | RELEVANCE TO CLINICAL PRACTICE

Healthcare professionals must act on the challenges that patients experience when deciding between non-invasive HMV and/or IHMV and their concerns and need for support in their decision-making processes. The knowledge that is provided in our study may increase awareness of the value of shared decision-making and patient decision aids so that patients and relatives can be involved in the best way and provided with greater knowledge of



treatment options and risks and resulting in patients and relatives experiencing less anxiety and fewer worries in their decision-making processes.

## AUTHOR CONTRIBUTIONS

All authors fulfil the four ICMJE requirements for authorship. Manuscript drafting and study design: Tina Thorborg; manuscript writing and analysis: Tina Thorborg; study conception, revision and approved the final manuscript: Jeanette FINDERUP, Charlotte Kirkegaard Lorenzen, Dorte Skriver Winther and Pia Dreyer.

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## CONFLICT OF INTEREST STATEMENT

The authors declare no conflicting interest in the research, authorship and/or publication of this article.

## DATA AVAILABILITY STATEMENT

The data set used and analysed in the current study is available from the first author TT upon reasonable request.

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