

## Accepting or declining non-invasive ventilation or gastrostomy in amyotrophic lateral sclerosis: patients' perspectives

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**Abstract** The objective was to identify factors associated with decisions made by patients with amyotrophic lateral sclerosis (ALS) to accept or decline non-invasive ventilation (NIV) and/or gastrostomy in a prospective population-based study. Twenty-one people with ALS, recruited from the South-East ALS Register who made an intervention decision during the study timeframe underwent a face-to-face in-depth interview, with or without their informal caregiver present. Sixteen had accepted an intervention (11 accepted gastrostomy, four accepted NIV and one accepted both interventions). Five patients had declined gastrostomy. Thematic analysis revealed three main themes: (1) patient-centric factors (including perceptions of control, acceptance and need, and aspects of fear); (2) external factors (including roles played by healthcare professionals, family, and information provision); and (3) the concept of time (including living in the moment and the notion of 'right thing, right time'). Many aspects of these factors were inter-related. Decision-making processes for the patients were found to be complex and multifaceted and

reinforce arguments for individualised (rather than 'algorithm-based') approaches to facilitating decision-making by people with ALS who require palliative interventions.

**Keywords** Amyotrophic lateral sclerosis · Non-invasive ventilation · Gastrostomy · Decision-making

### Introduction

Amyotrophic lateral sclerosis (ALS) is an incurable neurological disease characterised by progressive multisystem degeneration, with an average survival of 2–5 years after diagnosis [1–3]. Current interventions are limited to slowing disease progression or palliating symptoms such as dysphagia, weight loss, and respiratory muscle weakness [3, 4] and the recommended interventions, such as gastrostomy and non-invasive ventilation (NIV), tend to be required by people with ALS (pwALS) in later stages of the disease [3, 5]. Gastrostomy has been shown to improve nutrition, and NIV can help relieve the symptoms experienced due to diaphragmatic weakness, interventions which have improved prognosis and quality of life [6–9] without increasing caregiver burden [7]. Rates of uptake for NIV in the UK may currently be low [10]. While rates of acceptance of gastrostomy are not available for the UK, rates in the USA range from 0 to 63 % [9]. Evidence-based guidelines based largely on expert opinion recommend the use of NIV in ALS [11–14]. Findings [15] confirm observations from non-randomised trials and emphasise that NIV is part of 'best management' for people with ALS (pwALS).

The process of decision-making deserves exploration [16]. In quantitative studies, factors found to affect decision-making by pwALS in terms of NIV (and ventilation more generally) and gastrostomy include disease characteristics

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such as severity of symptoms [17]; demographic factors such as gender, age, educational level and IQ; and various psychological characteristics such as health beliefs, understanding of the illness, attachment to life, religiosity and mood (see [17–23]) as well as carer-rated everyday behavioural change indicative of executive dysfunction [17]. It has also been found that pwALS who indicate possible acceptance of NIV and gastrostomy early on in the disease trajectory are more likely to abide by their initial decision [24, 25]. In addition, acceptance of gastrostomy is related to a lack of pleasure obtained from oral intake and the inability to eat independently [26]. In a broader context, prospective research has demonstrated significant burden and psychological distress in caregivers of pwALS [7, 27, 28], but the relationship with palliative interventions has not always been explored [27].

Few qualitative studies have examined decision-making in ALS [29–31]. Research has elicited possible influences (including input from family and friends; religious and moral convictions and life sustenance [29]) and has identified themes such as the meaning of the intervention, the importance of context and values, the effect of fear, the need for information, and adaptation to or acceptance of the intervention [30]. Other influential factors were determined to be structural, interactional and personal [31]. It has been concluded [31] that decision-making is strengthened when patients experience co-operative relationships with healthcare professionals (HCPs) which support their personal beliefs. However, the findings are limited due to small sample sizes [29] and research has not specifically focused on gastrostomy and NIV [31, 32].

HCPs require sufficient knowledge to contribute effectively to decision-making [33] but there is mixed-quality evidence that non-ALS specialist HCPs lack knowledge of the disease [14]. Additionally, HCPs may have differing approaches to existing guidelines and conflicting opinions regarding the appropriate timing of an intervention [34] and discussions concerning NIV and gastrostomy are often initiated too late with little time for patients to discuss their preferences or for decisions to be unhurried [35, 36]. Nonetheless, the provision of information by HCPs has been found to be instrumental in patients' treatment decisions [37, 38].

Therefore, research exploring decision-making regarding interventions in ALS is mostly lacking. The qualitative studies that exist used semi-structured interviews or questionnaires [16, 39, 40] or investigated an a priori list of potential influencing factors [26]). However, they all provide limited understanding of the direct experience of pwALS in their specific decision-making process. The aim of the present prospective study was to identify from the perspective of pwALS other factors that influence decision-making, to understand better the processes involved in accepting or declining NIV and/or gastrostomy in ALS.

## Materials and methods

### Participants

PwALS were recruited from the South-East ALS register [41] as part of a larger, prospective study of decision-making about gastrostomy and NIV in ALS [17]. Inclusion criteria for this qualitative study were: confirmed diagnosis of ALS; duration of disease at entry into the larger study [17] between 6 and 60 months; and referral for NIV and/or gastrostomy, with a decision to either accept or decline the intervention. PwALS were not excluded on the basis of clinically apparent cognitive impairment, provided they could give valid informed consent. However, a detailed formal neuropsychological assessment of participants was not undertaken due to the number of measures administered in the larger study [17]; we had wanted to limit the burden on participants, both at entry to the study and following an intervention decision. At the latter time-point, patients' increasing ill-health may, in any case, have made it less likely that a lengthy neuropsychological assessment would have been acceptable to them.

Following an intervention decision, patients and their caregivers were invited to participate in a post-decision assessment including an in-depth interview. For those agreeing to an intervention, the assessment occurred after gastrostomy placement or NIV trial. For those refusing an intervention, interviews were arranged as soon as possible after decision confirmation.

### Ethics

Ethical approval was granted by the Joint South London and Maudsley and the Institute of Psychiatry NHS Research Ethics Committee (LREC 07/H0807/87). Written informed consent was obtained from all participants. The study was, therefore, performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments.

### Data collection

Semi-structured interviews were conducted by two experienced researchers (either by NHM—a clinical psychologist- or AJ, a post-doctoral researcher) in patients' homes and lasted 40–60 min. Those with speech difficulties used communication aids (e.g. a litewriter) or their carers spoke on their behalf. The interview schedule asked participants why they accepted/refused the intervention; their understanding of the intervention offered; their concerns related to interventions; their experiences leading up to making the decision including surrounding support and how the choice was offered. Open-ended questions were generated from a review of the literature and in

consultation with experts in the clinical management of pwALS and palliative care. The interview guide was amended iteratively and aimed to follow the priorities and concerns raised by participants.

### Analysis

Interviews were digitally recorded and transcribed verbatim. All identifying data were anonymized. An inductive thematic analysis approach [42] was adopted and data analysis software NVivo 9 (QSR International Version 9, 2010) was used to collate and manage the data. Initial transcripts were read repeatedly to enable the researcher (LPG) to become familiar with the data and identify preliminary patterns and themes. Data were divided into meaningful chunks and coded and the emergent themes were then examined for areas of convergence and divergence and grouped into higher level related concepts. A coding structure was devised and the ensuing framework was used as a basis for the remaining analysis. As new themes emerged in the residual data, the structure changed to incorporate developments and through this process of distillation, the main themes were produced. The superordinate themes were clustered within the main themes. The coding framework was regularly reviewed and refined through discussion between the research team (LPG, NHM, VL and LHG).

## Results

### Participants

From our larger sample of 78 people with ALS [17], a total of 32 people (41 %) made at least one intervention decision. Of these, 10 decided about both interventions. Our current sample comprised 21 people who agreed to and were well enough to participate in a post-decision interview (see Table 1). None of the 21 participants interviewed had declined NIV. Twelve participants were interviewed with their carers (these carers are denoted by ‘B’ in the tables) and a proportion of them communicated via a litewriter. One participant made two decisions and he and his carer (39A/B) were interviewed about each on separate occasions.

### Themes

Three superordinate themes emerged from the data that related to the decision-making process. These themes were: (1) patient-centric factors, which were directly related to the personal experience of the patient; (2) external factors, defined as influencing factors independent of the participant, namely the HCPs; his/her family and the issues surrounding information provision; and (3) the concept of time—the different aspects of time and their effects on the patient and his/her decision-making abilities.

**Table 1** Characteristics of patients taking part in the study and their intervention decision

Patient ID	Gender	Age at initial interview	Intervention	Declined/accepted
1A	M	68 years 4 m	Gastrostomy	Accepted
7A	M	71 years 9 m	NIV	Accepted
10A	M	68 years 1 m	Gastrostomy	Accepted
20A <sup>a</sup>	M	50 years 6 m	Gastrostomy	Accepted
27A <sup>a</sup>	F	66 years 2 m	Gastrostomy	Declined
30A	M	41 years 10 m	Gastrostomy	Accepted
33A	F	49 years 6 m	Gastrostomy	Declined
34A <sup>a</sup>	M	54 years 3 m	Gastrostomy	Declined
36A <sup>a</sup>	M	71 years 1 m	Gastrostomy	Declined
39A <sup>a</sup>	M	50 years 9 m	NIV and gastrostomy	Accepted both
42A <sup>a</sup>	F	72 years 1 m	Gastrostomy	Accepted
46A	F	62 years 4 m	Gastrostomy	Accepted
50A <sup>a</sup>	F	70 years 4 m	Gastrostomy	Accepted
51A <sup>a</sup>	M	65 years 9 m	Gastrostomy	Accepted
53A <sup>a</sup>	F	56 years 2 m	Gastrostomy	Declined
59A <sup>a</sup>	M	69 years 6 m	NIV	Accepted
63A	M	68 years 5 m	Gastrostomy	Accepted
64A	M	66 years 7 m	NIV	Accepted
65A <sup>a</sup>	M	52 years 4 m	Gastrostomy	Accepted
66A	F	76 years 2 m	Gastrostomy	Accepted
72A <sup>a</sup>	F	65 years 11 m	NIV	Accepted

<sup>a</sup> Participants interviewed with carers present

**Table 2** Evidence for themes—quotations from participants for patient-centric factors

Quote ref	Client ID	Accept/decline	Quote
<i>Perceptions of choice</i>			
2.1	50A	ACC	But I did not just sit there and fade away
2.2	53A	DEC	As far as I'm concerned it's my life, what's left
2.3	64A	ACC	And it can potentially prolong your life when even though all the MND symptoms carry on getting worse, you might have departed by then. [...] I don't want things keeping me alive when I'd frankly be better off dead
2.4	53B	DEC	Everyone was running around trying to make her change her mind and telling her about the benefits of having a PEG and that it would prolong her life for a little while, but she doesn't want it prolonged—53A always makes her own decisions, her brain still works, no one make her decisions for her
2.5	59A	ACC	It's not a choice, you either use it or you can't breathe of a night, so there is no choice
2.6	63A	ACC	If I'm not swallowing and can't eat anything and didn't have the PEG, I wouldn't survive. It's obvious I need an alternative method of feeding myself
<i>Acceptance and need</i>			
2.7	65A	ACC	My philosophy is to try to live as long as possible and I'm aware that MND patients die for two reasons: lack of nutrition and lack of air. So I was aware that I would need to tackle both of those things at some stage'
2.8	72B	ACC	If you think this is going to help you, you welcome it with open arms. You don't think 'well actually I don't want that' why would you? If it's going to help you, why would you not want it
2.9	64A	ACC	And that was the main thing I was aware that since there was an issue over my breathing, that was likely to get worse, so it wasn't just a question of 'I don't really need any help now' it was the knowledge that it was likely to continue to deteriorate, and if it would help me with sleeping and not having to get up so often.../
2.10	51A	ACC	Chewing food was getting more difficult, coughing, choking was getting more frequent
2.11	10A	ACC	Once I could see my throat was getting bad and I needed it, I could see it coming
2.12	42A	ACC	I don't really think about it, put it out of my mind
2.13	64A	ACC	No, no, no. I'd rather not know until there's a reason to know. I don't want to know all the nasty possibilities that might be in front of me. You know, I just don't want to know the detail, until there's a reason to know it
<i>Aspects of fear</i>			
2.14	33A	DEC	Yeh, it's the worry that it'll get infected
2.15	53A	DEC	What concerned me was the procedure
2.16	10A	ACC	I was a bit ... saddened... thinking about the strange thing coming out your stomach [...] I was thinking about my dad had cancer and he had a colostomy and it was thinking back to things like that..
2.17	27B	DEC	She was in hospital and they done a thing on her throat, [...] And she had trouble when she came round out of that. She had an awful lot of problems when they brought her round and I think that's frightened her of hospitals

A person with ALS, B carer of ALS patient

Of those interviewed, 16 agreed to the intervention and five declined. Data analysis aimed to explore similarities and differences in themes across these two groups.

Patient-centric factors (see Table 2)

Patient-centric factors emerged as influential in the decision-making process. These incorporated 'perceptions of choice and control', 'acceptance and need', and 'aspects of fear'.

#### *Perceptions of choice and control*

**Taking control:** The majority of participants took an active role in deciding whether or not to have an intervention. Whilst they could not control the diagnosis of a life-limiting disease, they wished to manage their lives and take active responsibility for themselves and their choices (see Table 2:

1 and 2), something participants commonly felt resided with them alone. Several pwALS appeared to be able to face up to the prospect of their own death (e.g., Table 2: 3). Remaining in control of one's body and not being swayed by others was also a strong theme (Table 2: 4).

**Absence of choice:** This theme was more prominent in the group who accepted an intervention because many viewed the intervention as an unavoidable consequence of ALS (Table 2: 5 and 6). Some participants expressed being unable to actively make a decision—the progression of the disease necessitated an intervention for them to continue living.

#### *Acceptance and need*

**Acceptance:** The acceptance of the condition and its subsequent progression emerged as a prominent theme among those accepting the intervention, with this also relating to

participants' acceptance of help. The decision was made because they wished to survive as long as possible and there would be a need for an intervention at some point along the disease's trajectory (Table 2: 7 and 8). Continuing deterioration was a trigger for some people that heightened their awareness of what the disease path was likely to be, and the impact of this on the patient was paramount in the decision-making process (Table 2: 9).

**Physical need for intervention:** This sub-theme was apparent largely in those who accepted an intervention. Reaching a physical stage (and being faced with clear symptoms) when participants believed there was no other choice, made the decision to accept the intervention more straightforward (Table 2: 10 and 11).

**Non-acceptance:** Some participants were forthright about their fear of the future. They were more focused on living in the present and only confronted issues when there was an obvious need (Table 2: 12). This meant that the ability to make a decision based on what could occur in the future was curtailed; they were in a state of active denial or non-acceptance (Table 2: 13). However, this did not seem to have an impact upon the final decision taken.

#### *Aspects of fear*

**Fear of the procedure:** The fear of the intervention itself was an immediate threat to some people and this influenced their treatment decision (Table 2: 14 and 15). Some had been advised to have a gastrostomy 'sooner rather than later', yet while they were still able to breathe or swallow/eat they found it difficult to view the intervention as necessary. Therefore, fear around the required operation was prominent whereas the timing of likely physical decline was unknown. There appeared, therefore, to be a direct relationship between fear of the intervention and it being declined.

**Prior health experience:** Some participants' decisions were coloured by a previous unpleasant health experience, either experienced personally or by someone close to them. This seemed to produce a strong fear response based around the prospect of repeating the upsetting event (Table 2: 16 and 17).

External factors (see Table 3)

The external factors that influenced acceptance or refusal of an intervention related to 'healthcare professionals' (HCPs); 'family' and 'information'.

*HCPs: Doctors, nurses, therapists, care workers*

**Supportive and supporting behaviours:** Decisions to have the interventions were taken with the support of HCPs and

participants' families. However, ultimately the decision lay with the patient. Some participants experienced their HCPs as being very supportive and caring, and were thankful that HCPs showed humanity and provided reassurance (Table 3: 1 and 2, respectively).

**Guidance:** Certain HCPs guided their patients into having an intervention (making 'the right decision') (Table 3: 3). Some informed patients about the consequences of leaving the intervention too late and were keen for their patient to have the procedure (Table 3: 4 and 5).

**Trust/lack of trust in the expert:** Directly related to 'Guidance', patients perceived that HCPs used their expertise to encourage acceptance of the intervention. Participants needed to feel they could trust their HCPs; some spoke about having no choice, as well as seeing the HCPs as needing to shoulder some of the responsibility (Table 3: 6). Participants also took HCPs' expertise seriously (Table 3: 7). Conversely, some were concerned by their apparent lack of knowledge about ALS (Table 3: 8) and felt under-supported in making a suitable decision.

**Decision pressure from HCPs:** Some participants felt pressurised by HCPs into making a decision (Table 3: 8 and 9), and experienced the frequent expression of the necessity of having an intervention as unpleasant (Table 3: 10). This highlighted tension around who was ultimately responsible for making the decision and who had enough information to 'know' what was the 'right thing'. Relevant here is the case of 20A (Table 3: 11), who fought pressure from HCPs to retain his own control. However, he eventually decided to have the procedure.

#### *Family*

**As a source of support:** Those pwALS who had support from their families appeared marginally more likely to agree to the intervention. Having supportive families played a major role in participants' ability to make a decision, feeling that the burden of the decision was shared (Table 3: 12).

**As emotional pressure:** Some patients experienced their families more negatively, with the family members' emotional needs taking precedence over those of the patient (Table 3: 13). Some decisions to accept an intervention evolved out of concerns for family members and not the participant's direct wishes (Table 3: 14).

#### *Information*

**Provision of information:** There was variability in how patients accessed information. Some sought to have this provided by HCPs and it was only in retrospect that they realised the paucity of what they had been told (Table 3: 15 and 16). However, others found that they were provided

**Table 3** Evidence for themes—quotations from participants for external factors

Quote ref	Client ID	Accept/decline	Quote
			HCPs
			<i>Supportive/supporting behaviours</i>
3.1	65A	ACC	'I think she didn't want us to make the decision, the final decision in her room. She wanted us to go back home and think about it
3.2	72A	ACC	I was concerned that it was something that would help me but could weaken my ability in the daytime. And it really reassured me that actually it would make it better. That was what I wanted to hear
			<i>Guidance</i>
3.3	63A	ACC	She explained whether I need it now or not
3.4	1A	ACC	His HCPs stressed the important of having the feeding tube early rather than when it's too late
3.5	33A	DEC	Have you thought about it because it would be a really good idea? And you should have it sooner rather than later. And his explanation is [...] that he believes that ultimately I will need one. He had an experience with patients before, who've left the decision to the last minute and then the operation's not gone well
			<i>Trust/lack of trust in the expert</i>
3.6	39A 39B	ACC	You're not a neurosurgeon or an MND specialist so you've got to be guided, you've got to trust that these people know what they're doing [...] We're lucky that we're young enough and sensible enough to be able to make decisions like that. But someone should be there to kind of take that responsibility away, we didn't go to university for 5 years and learn about medicine
3.7	33A	DEC	Well it made me think about it a bit more seriously than I would otherwise. [...] Clearly he knows more than I do about these things, and I was willing to listen seriously to it, and he mentioned it on two occasions so it was clearly something that he felt strongly that I should have
			<i>Decision pressure from HCP</i>
3.8	34B	DEC	They were like salesmen who didn't understand their product
3.9	66A	ACC	The main reason was that I felt persuaded into it
3.10	27A	DEC	I mean the speech therapist came round, she said 'well something you've got to start thinking about is this pipe' and [MND Specialist Nurse] came round and she said about it and that other woman said something about it an' all. It felt like a lot of pressure was being put on me
3.11	20A	ACC	The doctor tried to persuade me, my speech and language therapist has been trying to persuade me for more than two years. Every time he comes he nagged me, and we had an argument for nearly one hour, I was arguing that if it extends my life I don't want that
			Family
			<i>As support</i>
3.12	50A	ACC	[Our daughters] have made contact with the MNDA in Australia and they'd found out an awful lot about these things. And they were very keen on getting to it before leaving it too late. And I think having them here at the time sort of helped things along
			<i>AS emotional pressure</i>
3.13	66A	ACC	But my husband was clinging to a lifeline
3.14	53A	DEC	Of course my family are trying to keep me here—they don't want to hear of mum dying
			Information
			<i>Provision</i>
3.15	72A	ACC	The main phrase that sticks in my mind all the time is 'if you get to the stage where you need one of those ventilators would you have it?' and no, no more than that. And then you think, do I or don't I? Perhaps we should have asked more at the same time but...
3.16	36A	DEC	I think that if X, the MND nurse who was there, if she had said 'I've got some information, would you like to take it away with you just to have a look at?' that would have been quite useful
3.17	7A	ACC	I think I was given all the information I needed
			<i>Amount</i>
3.18	53A	DEC	Who explained that to you? Just everyone, doctors, nurses, matrons, everybody, loads of people
3.19	63A	ACC	I was given it all, so I knew about the disadvantages and the advantages
			<i>Actively seeking</i>
3.20	37A	DEC	The more information you have the more important decisions you can make, the more you know what the decisions are.
			<i>Accuracy</i>



**Table 3** continued

Quote ref	Client ID	Accept/decline	Quote
3.21	51A	ACC	If it had been clarified at the beginning, that there were two ways that you could have a feeding tube and the different ways that they're done and it could be that you will have one and it could be that you have the other, then that would have been helpful because then we could have looked them both up
3.22	33A	DEC	They certainly didn't go through anything about, you know, physical risks of the operation itself, and what might go wrong and why you should have it sooner rather than later so...
3.23	42B	ACC	No one said that she might feel sick, that came as a bit of a shock to her
3.24	65A	ACC	I had thought that once you had it, then almost everything would have to be sent down the PEG <i>Individualised information</i>
3.25	64A	ACC	There was more detail, but to be honest, I'm not that interested in the detail, just in the broader picture. If something can help, I'm not that interested in precisely the technicalities of how it helps, but just what benefit I can hopefully get from it and what the possible disadvantages are
3.26	72B	ACC	You can't take it all in at once anyway, so it's no good bombarding you with stuff from that point of view
3.27	34B	DEC	You said no to begin with but you wanted to keep checking that you were making the right decision and that's why he needed more information

A person with ALS; B carer of ALS patient

with enough information from various sources (Table 3: 17 and 18). Making the decision seemed to be easier for those who wished for and had access to different sources of information.

**Amount of information:** There was a small contingent who felt that they had been supplied with plenty of information and this, in turn, gave them confidence in making their decision [Table 3: 19).

**Actively seeking information:** Some participants took an active role in finding information; this bears direct relation to perceptions of control and responsibility. They sought advice to determine the best course of action. Those who decided against an intervention were more likely to actively seek answers to their questions (Table 3: 20).

**Accuracy of information:** An apparent lack of accuracy in the information offered to many participants caused consternation and disrupted the decision-making process (Table 3: 21). Participants reported a lack of consistency in information in relation to the disease and, in retrospect, what they should expect from the intervention (Table 3: 22 and 23). Some participants were less able to make a considered decision because they were unclear about aspects of the intervention (Table 3: 24).

**Individualised Information:** There was a clear need for information to be individualised. Each person had varying requirements and abilities and this added to the complexity of decision-making (Table 3: 25 and 26). According to his carer (34B), 34A (Table 3: 27) used his ability to reflect and analyse the information and choices available to challenge the HCPs, with this relating back to the themes of perception of control.

The concept of time (Table 4)

The concept of time was influential for the decision-making process and consisted of 'living in the moment'; 'right thing, right time' and 'predicting the future'.

*Living in the moment*

Some participants managed their situations by focusing on issues occurring in the present, their decisions emanating from that viewpoint (Table 4: 1), rather than worrying about the future (Table 4: 2). Some decided not have the intervention but were aware that as things changed, they might revisit that decision (Table 4: 3) signifying a fluidity within the decision-making process.

*'Right thing, right time'*

The idea of there being a 'right' moment for the 'right' decision was evident (Table 4: 4). Several participants had been advised to have the intervention whilst they were still physically strong enough to undergo the procedure but chose not to have the intervention then. However, due to the inherent variability in disease progression, it was difficult for patients to know how, and when, to make a decision, as this would only be clear in retrospect (Table 4: 5). Not being able to predict the course of the disease made decision-making complicated (Table 4: 6 and 7). Decisions had to be made based on what was happening at the time, as well as what could happen in the future (Table 4: 8). Comments by 37B (Table 4: 9) question the language that HCPs used to assist patients in making decisions—using

**Table 4** Evidence for themes—quotations from participants for the concept of time

Quote ref	Client ID	Accept/decline	
4.1	39B	ACC	Until you get to that stage you don't actually focus on it because you're actually focusing on the stage where you're on. And that kind of consumes all your energy and all your time. So until you actually reach that point you don't deal with it
4.2	72A	ACC	We're dealing with things as we have to, and it's probably not a bad way of going about it. And just thinking I'll worry about it when it gets here. I can't worry about it before
4.3	33A	DEC	So I'm just going to wait for that stage before I review the decision
4.4	65A	ACC	It was very much the timing, which was what we were thinking about, rather than actually doing it or not
4.5	33A	DEC	So I'm not saying never, but I don't want to decide yet, so my decision now is 'not at the moment'
4.6	39B	ACC	The problem with these feeding tubes are that you expect the professional people to guide you, tell you when's the right time but they don't
4.7	39B	ACC	You can't make a judgment because you don't know what you're judging really
4.8	59B	ACC	Well, it was too quick to know, you know what I mean? You don't know how this disease is coming on, you don't know what's next
4.9	37B	DEC	They say make sure it's done sooner rather than later but what is sooner rather than later? They don't say we're talking next month, no definite time, it's down to you

A person with ALS, B carer of ALS patient

for example a 'stock phrase' (e.g. "sooner rather than later") was perceived as unhelpful to patients, as it highlighted a lack of knowledge of their likely disease trajectory.

## Discussion

This study investigated the factors involved in decision-making regarding acceptance or refusal specifically of NIV and/or gastrostomy for pwALS in a prospective UK sample drawn from a population register. Relative to recent qualitative studies [e.g., 31], the sample was large and all participants took part in face-to-face interviews. Three main themes emerged from the analysis: (1) patient-centric factors which included perceptions of control, acceptance and need, and aspects of fear; (2) external factors which included the roles played by healthcare professionals, family, and information; and (3) the concept of time, including living in the moment, and the notion of 'right thing, right time'. Many of these factors were inter-related and decision-making processes for pwALS were found to be complex, multifaceted and individual. The patient-centric factors had the most immediate and direct relationship with the decision-making process, followed by external factors (wherein the support or lack thereof and the pressure to make a decision from the HCPs and family members had varying impacts); here, the amount and quality of information given or sought were also important. Our findings highlight that while pwALS are respectful of HCPs' expertise and knowledge, they recognise when HCPs are less

well informed about interventions and this may have a negative impact on patients' experience of decision-making. Finally, the concept of time served as an underlying influence.

Our UK-based findings are largely in line with previous studies (both quantitative and qualitative) on decision-making in ALS [e.g. 17–23, 29–31]. As elsewhere [31], the relationship between pwALS and their HCPs was important in decision-making, and the support, or lack thereof, offered by HCPs had a direct impact on whether or not an intervention was accepted. There was a clear discrepancy between pwALS whose priorities were more focused on their current situation, as discussed elsewhere [31], and HCPs' 'best practice' suggestions of interventions to counter future difficulties related to the progressive nature of the disease. This 'worst-case' decision-making context [31] was experienced by some pwALS as pressure, which added to their distress. It is possible that during the course of this study such pressure may have increased due to the publication of UK guidelines regarding NIV provision [14], which may have resulted in an increase in the number of pwALS being offered NIV. In contrast, other pwALS felt supported and understood by their HCPs.

The role of informal carers in decision-making by pwALS has been interpreted as enhancing ALS patient-centred care [31] by contributing to a patient-carer-HCP decision-making triad; the challenges of this relationship were not fully explored in the current study. Nonetheless, our findings suggest that while supportive carers may facilitate acceptance of interventions, they may also contribute to pressure on pwALS to opt for an intervention



counter to their own preferences. Elsewhere, it has been suggested that it may be important for pwALS that their families accept their care-related decisions [32], while those with no immediate family may feel less constrained in the decisions they make.

Another important factor in the decision-making process was whether pwALS perceived that control over their illness resided with them or with HCPs and the interventions they could provide. Those who felt they had no choice but to accept an intervention considered the disease was dictating the need; they appeared to have an external health locus of control (i.e., they believe they or others have the ability to control their health status) in contrast to those who wished to maintain control over the disease itself and not rely on external interventions. Evidence of physical deterioration signalled a need for an intervention to sustain life; this was a strong factor influencing acceptance of an intervention and was consistent with previous studies [19, 26]. Our findings, as elsewhere [32], also indicated that pwALS' prior medical experiences might need to be explored and understood when advice is given about interventions.

The theme 'the concept of time' was complex and, from the perspective of pwALS, was experienced differently by them and their HCPs. For the pwALS, 'time' was experienced as something relating either to the present or the past. There was a sense that pwALS here, as in other studies [31] were choosing to focus on the immediate and 'known', and not to contemplate the future. This may have been due to their inability to determine the specific future trajectory of the disease as well as being in a state of denial or non-acceptance, and electing to focus attention on aspects of their condition over which they felt they currently had some control. In contrast, pwALS felt that the HCPs who tended to view time as 'lacking' placed pressure on pwALS to have an intervention. In reality, the unpredictable nature of the disease course means that there may need to be a cyclical pattern of decision-making as symptoms change [43] and life-stage transitions may also determine the decisions likely to be made [32]. This finding may help HCPs to develop more patient-focused ways of supporting and advising pwALS.

Current findings concur with results from a quantitative study on the same participants [17], where those people who were more active in their information-seeking were more likely to refuse an intervention. This may be related to specific individual characteristics and coping styles but it is clear that receipt of more information did not necessarily result in acceptance of an intervention. This may be due to greater understanding of the implications or side effects of an intervention, resulting in pwALS deciding to retain the 'status quo'. Our related study [17] also found that those with less understanding of their illness were also more

likely to refuse an intervention. This may be related to fear or misguided perceptions about the intervention or could more generally reflect a lack of, or poor quality, information. Certainly, those who were specifically concerned about having the procedure were more likely to decline an intervention, suggesting a relationship between information-seeking behaviours, quality and accuracy of information sought or offered and understanding.

Interestingly, our current study did not reveal any evidence for influences of spiritual determinants or religiosity on decision-making that have been found in other papers [23, 29] and echoes other findings [31]. However, the role of religiosity/spirituality in decision-making was not specifically asked about and this could be seen as a limitation: religiosity (found elsewhere to maintain quality of life in pwALS [44]) may be something that is only discussed once prompted. In our related quantitative study where pwALS were specifically asked about religiosity, lower levels of religiosity were more likely to be associated with intervention refusal [17].

As noted earlier, participants did not undergo formal detailed neuropsychological testing in this study, nor did we explore the role of cognitive involvement [45] in decision-making in our interviews. Of relevance, however, is that our related quantitative study [17] indicated that the presence of everyday executive dysfunction in patients (as rated by caregivers) at study entry and ratings of pre-illness executive dysfunction and disinhibition were likely to be associated with the subsequent refusal of an intervention. That study [17] questioned whether more impaired patients might be less able to evaluate information about interventions and indeed whether they are in some implicit way discouraged from undergoing interventions, through concern over compliance in cognitively impaired patients [46]. Our wider findings [17] resonate with other work suggesting that cognitive impairment might potentially reduce patients' involvement in decision-making [31], and might argue for at least routine screening (if not full neuropsychological assessment) of cognitive and behavioural change [e.g., 47] when considering patients' ability to engage in decision-making.

## Implications

Importantly, those who had declined interventions felt they had the choice—at a later stage in the disease's trajectory—to rescind their decision. There seems to be a pattern of cyclic revisiting as the situation for pwALS is one of change [43]. This study captured the decision-making process at the point at which a decision was made. However, pwALS may well have been considering options for interventions over a longer period of time and more enduring psychological and emotional characteristics may be

explanatory factors [17]. Whilst there is a definite moment at which a decision is communicated it can take variable lengths of time for the person to reach that point. Carers and HCPs need to be attuned to the dynamic nature of this process.

This paper highlights the need to offer pwALS individualised support to aid decision-making. For example, being able to recognise those who have an ‘active’ approach as opposed to those who are more passive (in such things as information-seeking) and developing individualised support may be beneficial to patients’ decision-making. Simply responding to treatment guidelines may lead to pwALS perceiving themselves as being pressurised. Conversely, recognising that those who took a more active approach to information-seeking were ultimately more likely to refuse an intervention should not result in HCPs being less willing to address patients’ concerns about an intervention.

Those pwALS who believe that the choice to accept or decline intervention is ultimately theirs appear to respond differently from those who feel that the control is external to them and that they are being controlled by the disease and supported (or not supported) by the HCPs responsible for their care. It may be difficult for an HCP to gauge the correct level and depth of information for their patient, depending on the patient’s health locus of control. Offering clear information on quality of life and symptomology post-intervention, as well as taking into account levels of fear, may go some way to ease the decision-making process.

### Limitations

As with all qualitative methodology, this study’s findings may only generalise to the population from which this sample was drawn, and only represent the specific questions asked in the interviews. However, the involvement of pwALS drawn from a population register may overcome some potential limitations of clinic-based studies.

Due to communication difficulties, as in other studies (e.g. [32]), some pwALS’ carers spoke on their behalf; hence whilst the views may well have been those of the pwALS, they would have been shared through the filter of the carer. However, we considered it more important not to exclude pwALS with communication difficulties.

In this study, only five pwALS were interviewed who had declined gastrostomy and thus reasons for refusal could not be explored as thoroughly. Alternatively, this could indicate that proportionately more pwALS opt to accept this intervention rather than decline. In a similar way, there was a lack of interviews for those who declined NIV. This, however, may be due to the fact that NIV is usually required during the later disease stages [3]; hence, pwALS

are generally more unwell at this point. In this study, those who declined NIV died soon after making their decision [17].

In conclusion, this study suggests that decision-making for intervention in terms of NIV or gastrostomy in ALS is a complex, unpredictable and fluid process, influenced by relationships with HCPs, with family and related to the concept of time. The discrepancy between pwALS’ autonomy and HCPs’ guidance towards intervention (experienced by some as pressure) demonstrates the need for individualised treatment and for more patient-focused support.

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