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Why don't they accept non-invasive ventilation?: Insight into the interpersonal perspectives of patients with motor neurone disease

Hikari Ando^{1,2,3*}, Carl Williams¹, Robert M. Angus²,
Everard W. Thornton⁴, Biswajit Chakrabarti², Rosanna Cousins¹,
Lucy H. Piggin⁴ and Carolyn A. Young³

¹Department of Psychology, Liverpool Hope University, UK

²Chest Centre, Aintree University Hospital, Liverpool, UK

³The Walton Centre NHS Foundation Trust, Liverpool, UK

⁴School of Psychology, University of Liverpool, UK

Objectives. Although non-invasive ventilation (NIV) can benefit survival and quality of life, it is rejected by a substantial proportion of people with motor neurone disease (MND). The aim of this study was to understand why some MND patients decline or withdraw from NIV.

Method. Nine patients with MND (male = 7, mean age = 67 years) participated in this study. These patients, from a cohort of 35 patients who were offered NIV treatment to support respiratory muscle weakness, did not participate in NIV treatment when it was clinically appropriate. Semi-structured interviews and interpretative phenomenological analysis (IPA) were employed to explore these patient's experience of MND and their thoughts and understanding of NIV treatment.

Results. Using IPA, four themes were identified: preservation of the self, negative perceptions of NIV, negative experience with health care services, and not needing NIV. Further analysis identified the fundamental issue to be the maintenance of perceived self, which was interpreted to consist of the sense of autonomy, dignity, and quality of life.

Conclusions. The findings indicate psychological reasons for disengagement with NIV. The threat to the self, the sense of loss of control, and negative views of NIV resulting from anxiety were more important to these patients than prolonging life in its current form. These findings suggest the importance of understanding the psychological dimension involved in decision-making regarding uptake of NIV and a need for sensitive holistic evaluation if NIV is declined.

Statement of contribution

What is already known on this subject?

- Non-invasive ventilation is widely used as an effective symptomatic therapy in MND, yet about a third of patients decline the treatment.

*Correspondence should be addressed to Hikari Ando, Neuroscience Research Centre, The Walton Centre NHS Foundation Trust, Lower Lane, Fazakerley, Liverpool L9 7LJ, UK (email: hikari.ando@nhs.net).

- Psychological disturbance generated by NIV use leads to negative experiences of the treatment.
- Decision-making about treatment potentials is complex and unique to each individual affected by perceived impact of disease.

What does this study add?

- A decision concerning NIV uptake was influenced by perceived impact on individuals' sense of self.
- Sense of self was influenced by the maintenance of autonomy, dignity, and quality of life.
- Individuals' sense of self was identified to have been challenged by the disease, NIV, and their experience of health care service.

Motor neurone disease (MND), also known as amyotrophic lateral sclerosis (ALS), is a fatal neurodegenerative disease indicative of the loss of upper motor neurons (UMN) and lower motor neurons (LMN; Kiernan *et al.*, 2011). The clinical features of UMN are moderate weakness, hyperreflexia, pathological reflexes, pseudobulbar affect (emotional lability; sudden and uncontrollable episodes of crying and/or laughing), spasticity, loss of dexterity with hands, and slowed movements (Strong & Rosenfeld, 2003). The clinical features for LMN involve severe weakness, hyporeflexia, muscle atrophy, fasciculations, muscle cramps, and decreased muscle tone (Strong & Rosenfeld, 2003). Although MND is primarily a disease of motor system degeneration, it is now recognized that there are overlaps between MND and frontotemporal dementia from pathological, clinical, and genetic perspectives (Neumann *et al.*, 2006; Phukan, Pender, & Hardiman, 2007). In the overall MND population, about 50% of patients die within 30 months of the onset and about 20% of patients survive between 5 and 10 years after the symptom onset (Talbot, 2009).

There is currently no cure for MND; however, various symptomatic therapies are useful and are offered to patients. The domain of potential therapies includes educational, pharmacological, and psychological interventions and the supply of medical equipment, such as non-invasive ventilation (NIV). Addressing respiratory issues is important as most deaths in MND are caused by pulmonary complications (Andrews, 2009; Winhammar, Rowe, Henderson, & Kierman, 2005). NIV is the treatment of choice to assist respiratory effort, and it increases life span, however, with little survival benefit for individuals with severe bulbar impairment (Bourke *et al.*, 2006). NIV is widely recommended as a standard treatment (Andrews, 2009; Phukan & Hardiman, 2009), and the recent survey in the United Kingdom showed an increased number of patients with MND using NIV (O'Neil *et al.*, 2012).

Non-invasive ventilation is also widely recognized to improve patients' quality of life (Bourke *et al.*, 2006; Sundling, Ekman, Weinberg, & Klefbeck, 2009). However, 'quality of life' is a generalized concept, and there is a lack of consensus on its component parts (Cella, 1994). It is often measured quantitatively thus falling short of reflecting a detailed idiographic account, especially of those categorized as 'uncommon'. This becomes a particularly important concept where patients do not take up NIV, raising the question: 'What are the dimensions of quality of life of those who decline a treatment known to be beneficial?' This study reports an investigation into the experience of those who decline the treatment towards achieving a better understanding of the full impact of NIV treatment in MND.

Experiences of NIV

Previous studies of NIV have mainly focused on benefits of the treatment, and these benefits are often measured by means of comparison between NIV users and non-users

(Lo Coco *et al.*, 2006; Mustfa *et al.*, 2006; Sivak, Shefner, Mitsumoto, & Taft, 2001). While these studies acknowledge that NIV is not always tolerated or accepted, little attention has been paid to exploring the factors, leading up to the decision to use NIV. While there is some evidence that a supportive caregiver is required for uptake of NIV treatment (Cousins *et al.*, 2013) to date, there remains a lack of a full account of reasons for not accepting NIV treatment. Moreover, this is particularly of interest where individuals have initially been willing to use NIV for their condition, yet later decline, as it was for some of the participants in the current study.

Although previous studies do not provide sufficient reasons for declining or withdrawing from NIV, unfavourable features of NIV have been identified by studies exploring the experience of people already established with NIV (Lemoignan & Ells, 2010; Lindahl, Sandman, & Rasmussen, 2005). Studies exploring the experience of NIV in various conditions, including MND, seem to agree that both a burden and a relief are associated with NIV usage. Some of the reported negative experiences of NIV are feelings of being in a vacuum cleaner bag, being exposed to air pollution, and following the rhythm of NIV rather than the machine following the individual's breathing pattern (Lindahl *et al.*, 2005). One prominent feature among the negative aspects of NIV is the mask (Lindahl, Sandman, & Rasmussen, 2003; Torheim & Gjengedal, 2010). It has been expressed that the mask can intensify anxiety by giving a sense of being trapped in it. This sensation negatively affects breathing which, in turn, causes a sense of losing control over the situation, inducing panic and irrational behaviour (Torheim & Gjengedal, 2010). In addition, the foreign nature of the mask creates a sense of otherness (Lindahl *et al.*, 2003; Torheim & Gjengedal, 2010), indicating an escalated threat to their identity. Generally, however, it is perceived that the burden of NIV is primarily a result of the psychological disturbance generated by the implications of their need for a breathing assistance device and the fear of being dependent on it: existential and independence threats (Lemoignan & Ells, 2010; Lindahl *et al.*, 2005; Lindahl, Sandman, & Rasmussen, 2006).

Decision-making

In contrast to the previous paternalistic approach in medical intervention, a decision over preference of care, including life sustaining treatment, is a joint decision between clinicians and patients with the latter being the ultimate decision-maker in current Western society. Gauthier and Swigart (2003) observed the decision-making in terminally ill patients and report three determinants in decision-making process: *realization of terminality*, *accommodating living*, and *engagement with uncertainty*. The first factor refers to the degree of awareness of mortality that involves reflection on physical ability and admitting to the changes. The second involves the realization of their needs and adaptation to the changes. The last involves preparing for the unknown future with hope, preparation, and anticipation. These three concepts were found to interact with each other and work as assessment criteria to understand where individuals stand in the decision-making setting. This model fits in with the findings of previous studies. For example, Reid and her colleagues found that patients with cancer viewed opioid treatment as the last straw and were resistant to it until the symptoms became too severe to endure (Reid, Gooberman-Hill, & Hanks, 2008). This attitude towards opioids may be understood as a wrestle between the acceptance of death and physical deterioration (*realization of terminality*): an attempt to deny mortality by not taking opioids as this is generally associated with being in the terminal phase. Johnson and Nelson (2008) also observed denial of the reality to negatively affect decision-making by the dismissing of

treatments, further supporting the model. And although there is a need for more investigations to explore this argument, the importance of having a purpose in life as a motivation for life (Chapple, Ziebland, McPherson, & Herxheimer, 2006) may be explained using this model; the future without a particular aim, or simply a lack of hope for the future in the face of awareness of negative physical changes, is evaluated as pointless. Such an attitude towards the future may be affected by a reluctance to adapt to changes caused by disease, resulting in failing to generate adapted goals for the altered life.

In addition to the criteria proposed by Gauthier and Swigart (2003), the sense of control and perceived quality of life have been recognized as other influential factors in how people make decisions (Chapple *et al.*, 2006; Lipsman, Skanda, Kimmelman, & Bernstein, 2007). Chapple *et al.* (2006) argue that individuals may choose to actively welcome euthanasia as a way of controlling their lives, while losing the control over their body, and the right to make such a decision is seen as preserving one's dignity.

Similarly, the decision regarding NIV use among people with MND is likely to be dependent on their perception and evaluation of worthwhileness of their existence, alongside a consideration of their physical deterioration and its psychological consequences. Previous studies indicate that the decision-making process is a complex procedure involving individuals' evaluations of their physical condition, acceptance and adaptation to the changes, and their sense of control and quality of life.

Current study

The aim of this research was to find the underlying reasons for why 30% of those offered NIV treatment in our main study declined or withdrew from the treatment, and to gain an in-depth understanding of a patient's experience and decision-making processes around an offer of NIV as part of treatment of MND.

Method

This investigation is a part of a larger prospective study (Ando *et al.*, 2014; Cousins *et al.*, 2013). The aim of the larger study was to explore the experience of NIV treatment over time in patients with MND and their caregivers, which is being undertaken at two National Health Service foundation trusts in North West England, United Kingdom. The study was designed as a longitudinal study where patients and their caregivers were followed up every 3 months from the time of referral to respiratory assessment until their death, or withdrawal from research. All patients knew that the purpose of referral was to commence respiratory monitoring in case NIV was required in future, and were aware of the potential to use NIV in MND care. The larger study employed mixed methods, and the current article presents findings from the qualitative phase of the study.

Ethical concerns

All potential participants were given an information sheet providing full details of the study, and assuring confidentiality and anonymity. Their right to decline or withdraw from the study was explained, and it was emphasized that there would be no negative effect on their treatment as a consequence of their decision. Consent was obtained once patients agreed to take part. This study was approved by an NHS Local Research Ethics Committee.

Participants

Inclusion criterion for the main study was a confirmed diagnosis of MND based on El Escorial criteria (Brooks, 1994); exclusion criterion was the presence of symptoms indicating cognitive or behavioural dysfunction. NICE (2010) guidance was used for both the timing of the referral to further respiratory assessments and for NIV initiation. Potential participants were identified by the respiratory team when they were referred for respiratory assessments.

Thirty-five patients were enrolled into the main study. 10 patients did not take up NIV treatment when it was offered based on physiological markers. One patient was subsequently excluded from the analysis as the patient declined to talk in-depth about the illness, limiting the sufficiency of the data. Of the remaining nine patients who provided data for this research, seven patients undertook an NIV trial when it was offered; six patients decided fairly quickly to withdraw from NIV; one patient (Henry) used NIV for 2 months before withdrawal from the treatment. Two patients did not try NIV treatment, and both declined their respiratory follow-up appointment. Table 1 provides both demographic and clinical information and data on the basis of recommendation made for an initiation of NIV.

Data collection

Semi-structured interviews were employed to allow the researchers to be flexible in exploring any interesting and relevant issues mentioned by participants (Smith & Osborn, 2008). The interview schedule for the main study was developed by a multidisciplinary team that consisted of one neurologist, two respiratory physicians, and one psychologist. The interview schedule included exploring personal views of NIV treatment and participants' perception of the impact of NIV treatment over time. The first interview was before trialling NIV; participants' impressions, knowledge, and perceptions of both MND and NIV (e.g., 'When NIV was offered for your breathing, what were your thoughts?') were explored. Post-trial interviews then investigated their experience of NIV and their decision to withdraw (e.g., 'What was it like to use the ventilator?'). The length of interviews varied between 20 min and 1 hr. The interviews were conducted at either at the patient's home or in hospital, according to the patient's preference. Patients were being interviewed on a regular basis by two researchers for the main study. The

Table 1. Demographic and baseline medical characteristics of the participants ($n = 9$)

Patient	Age	Onset	Bulbar at the time of trial	Basis of non-invasive ventilation recommendation
John	58	Bulbar	Yes	Nocturnal hypoventilation*
Matthew	79	Bulbar	Yes	Nocturnal hypoventilation*
Simon	71	Bulbar	Yes	Shortness of breath, Orthopnoea
Catherine	64	Limb	Yes	Nocturnal hypoventilation*
Henry	77	Limb	Yes	Nocturnal hypoventilation*
Peter	72	Limb	Yes	Mild breathlessness
Ian	71	Limb	No	Nocturnal hypoventilation*
Rachel	40	Limb	No	Nocturnal hypoventilation*
Ted	71	Limb	No	Orthopnoea

Note. *Measured by nocturnal pulse oximeter.

researchers were known to be independent of the clinical care teams. Participants' preference regarding the presence of their caregiver during their interview was respected, although it was preferred to interview the person alone.

Interviews were normally conducted verbally. However, some strategies were employed to mitigate communication difficulties with patients who had dysarthria (e.g., communication by writing). The need for communication alternatives has been previously argued and justified (Philpin, Jordan, & Warring, 2005). Interviews were audio recorded and transcribed verbatim except for interviews with John who did not want to be recorded, and therefore, only field notes were available. This approach of recording field notes is highly recognized in ethnography, where field notes may be the sole accessible data (Palmer, 2010). In the same aspect, field notes were used in the current research as the only data for John, who declined the formal interview, yet remained keen to participate in the study. Although it may be argued that the analyses of John's interviews, or those with communication difficulties, should be excluded, the limited number of people who fit into this sample category should be recalled. It was essential to hear the views of this particular group of people on NIV treatment, albeit in this case, through field notes rather than a verbatim transcript.

Pre-trial interviews were conducted with all nine participants; we were only able to conduct post-trial interviews with five patients due to death or withdrawal from the study. The first post-trial interviews were used for analysis with five patients. Post-trial interviews were not available for four participants: two withdrew from the NIV trial and two withdrew from both the trial and the respiratory service. The closest interview to the time of trial, or declining from the trial, was analysed for those without a post-trial interview.

For Simon and Ian, two interviews were used as there was a change in interviewers between those interviews. Including an additional interview in the analysis for these two patients was decided upon in consideration of the need to fully explore those participant's experience during the imperative period of time. Two interviews were also analysed to better understand reasons behind declining NIV for John as only field notes were available. Therefore, 5 pre-NIV interviews and 7 post-NIV interviews were available giving a total of 12 interviews for analysis. Medical data in relation to interviews and the timing of the interviews are listed in Table 2. Although participants were recruited at the time of consideration for NIV, the trial of NIV depended on results of their respiratory assessments, and this resulted in variance in timing of interview visits.

Data analysis

As the aim of the study was to understand underlying reasons for unwillingness to use NIV, an ideographic and phenomenological approach was considered most appropriate method. Interpretative phenomenological analysis (IPA) was employed as it aims to understand a particular phenomenon experienced by a particular group of people, yet without losing the particularity of each participant (Smith, Flowers, & Larkin, 2009). Through detailed examination of the phenomenon, as explained by the individuals, IPA strives to identify the significance of the experience for the individuals, and this often involves a small cohort (Brocki & Wearden, 2006; Smith *et al.*, 2009).

The analytic steps described by Smith *et al.* (2009) were followed in the current study. The analysis involved familiarization with the data, comprehensive annotation of initial notes on the transcripts, and development of emergent themes. The themes were arranged according to their relevance to the aims of the research and to their significance in relation to reflecting participants' lived experience of MND and NIV. A

Table 2. Medical data in relation to interviews and NIV trial

	Patient	Duration of MND before NIV trial (month)	Duration of MND before interview(s) (month)
Post-NIV trial interviews available	Henry	1	3
	Ian	27	25, 28
	Matthew	17	17
	Rachel	1	4
	Ted	118	120
Deceased without post-NIV trial interview	Catherine	14	12
	Simon	16	10, 15
Declined from NIV trial	John	–	24, 28
	Peter	–	20

Note. MND, motor neurone disease; NIV, non-invasive ventilation.

theme table was created containing both super-ordinate themes and their subthemes. Individual analyses were first carried out, followed by broader generalizations of the reasons for declining NIV treatment, in the context of MND, through exploring the patterns across the cases.

Although the analysis was conducted by one researcher, rigour of the analysis was achieved through peer examination (Krefting, 1991) with two other authors at two separate points: at an early stage of the analysis and at the time of creating the theme table. The peer examinations tested the credibility of interpretation of the quotes. The analysis is also made transparent by the use of verbatim quotes in order that the credibility of the analysis may be examined. It should be understood, however, that the interpretation presented here is not the sole interpretation of the accounts.

Findings

A summary of the analysis is illustrated in Table 3. Four superordinate themes were derived from patient narratives: preservation of the self, negative perceptions of NIV, negative experience with health care services, and not needing NIV. These are elaborated below with each of these themes described with an indication of their meaning for the patient and illustrated by verbatim quotes from their narratives.

Preservation of the self: 'it's not really me anymore. . .'

Physical changes in MND were always mentioned by all of the participants as unavoidable phenomena; however, the consequences of the changes were especially referred to by four participants:

And as well, she didn't shave me so I have no shave now and I hate that. I've always had a shave every day of my life I have. I think that might be the worst thing about this motor neurones. (Matthew, post-trial)

When talking about his caregiver, Matthew expressed his annoyance with her not performing his previous ritual of 'shaving'. Albeit a simple change in his daily activity, this

Table 3. Overview of analysis themes

Superordinate theme	Themes	Subthemes	Participants
Preservation of the self	Identity	<i>Damaged identity</i>	Catherine, Matthew, Simon
	Preservation	<i>Autonomy</i>	Catherine, Matthew, Simon
		<i>Dignity</i> <i>Quality of life</i>	Catherine, Ian Ian, Simon
	Breathing	<i>Significance of breathing</i> <i>Autonomy of breathing</i>	Catherine, Simon Catherine, Simon
Negative perceptions of non-invasive ventilation (NIV)	Features of NIV	<i>Mask</i>	
		Sound	Catherine, Simon
		Alien	Ian
		Appearance and practicality	Matthew
		Claustrophobic	Ian, Ted
		<i>Air Pressure</i>	Matthew, Ted, Rachel
		Shocking	
	Psychological reaction to NIV	<i>Fear</i>	Matthew, Ted
		<i>Unbearable</i>	Ian
	Threats to autonomy	<i>The loss of control</i>	Ian, Matthew, Ted, Rachel
Negative experience with health care services	Impression of hospital	<i>Dislike of hospital</i> <i>Poor image of hospital</i>	Ian, Matthew, Ted Ian
	Bad interactions with hospitals and their anticipated outcomes	<i>Emotional damage</i>	
		Withdrawal	John, Peter
		Rejection	Peter
		Confusion	Ted
Not needing NIV	Symptoms and perceived cause	<i>Disempowerment</i>	
		Invaded autonomy	Ian, Simon
		<i>Symptoms observed</i>	
		Breathing	Catherine, Ian, Matthew, Simon
		Sleep	Ted, Rachel
		<i>Perceived cause</i>	
		Different causations	Catherine, Ian, Matthew, Ted, Rachel
		Chance	Matthew, Ian
The need for NIV		<i>Solvable changes</i>	Catherine, Simon
		<i>Suspicious about the need</i>	Catherine, Ian, John, Matthew, Ted, Rachel, Simon
		<i>Terminal phase of the illness</i>	Henry

was perceived to be critical. This notion of curtailment of previous habits was further linked with changed identity as noted by Simon:

I always used to wear a shirt – had to. You know, had to be smart, take care, look good. Always used to wear a shirt every day and have a shave and now I can't. I have to wear tops without buttons and I hate that. It's not really me anymore because of what I can't do – the fiddly things

like buttons. In business, it's important, you know, dress the part – not like this, I hate this now that I'm wearing. (Simon, pre-trial)

Even though his habit of wearing a shirt was described as compulsory, nevertheless, wearing a shirt was a recognized part of Simon's identity, which was now damaged because of his physical restriction. The shirt was understood to be a 'part' of being a businessman; thus, his previous habit of wearing a shirt was a crucial attachment to his identity. Despite the fact that Simon was no longer working in business, he still desired to retain the previous normality, yet his physical impairments challenged this. This struggle between their past and the new identity was a common observation, and the new identity was generally perceived to be unacceptable:

I don't want to just [long pause] fade away, just a helpless thing. That really is just the worst thing that could ever happen to me; because, well, it must be hard for you to see it now because you never knew what I was really like. But it's not how I've lived my life, like this, and I'm just so sure that it's not how I want to go out either. I want this all really to be over as soon as it can be. I wouldn't want to live much longer, and certainly I wouldn't want people trying to keep me alive longer than was natural – Longer than the disease wanted me alive. So, if there's no cure, I don't want to be messing with anything else, you know. (Catherine, pre-trial)

Catherine described how MND had damaged her life. MND had made her into a different person distinct from her previous self. She questioned whether her impaired autonomy diminished her quality as a human being. Her devastation over her current condition was expressed in her depersonalization of herself as a 'helpless thing'. It can also be interpreted that the process of 'fading' of her physical body is also causing the 'fading' of her sense of existence as a human. Catherine had little interest in 'messing with anything' in this context, which indicates her prioritizing the preservation of her dignity and her sense of self. The concern for dignity was prominent for all participants, and it related both to the way of maintaining and of ending their lives. Ian uses an example of another patient with MND to portray this point:

What's-his-name, um, [MND patient] well, but look at the state he's in. What kind of a life is that? I mean his brain is still alright; that's still alive, but that's all though. (Ian, pre-trial)

Ian seemed to be suggesting that the quality of being consists largely in mobility, and it may be further understood that the mere functioning of the brain without any mobility does not count as a life as such, but just an existence. This points to the importance of perceived quality of life, and mobility was the important aspect of quality of life for Ian. The image of lifeless existence with the illness, either with or without cognitive functioning, was also raised by other patients, and NIV was perceived to be a symbol of such an existence.

I don't want to end up like one of those people who just lives hooked up to a machine. They're not really living, I wouldn't want it to be all of the time and not have any life. When I first heard it [NIV] – that was what I thought. That was my first thought when I heard it. That's what I'm afraid of – if I start on that a little bit. [Long pause] I don't want that. You can't do anything. (Simon, pre-trial)

Non-invasive ventilation presented an obstacle to having a life, and Simon claims that life cannot be obtained while 'hooked up' to a machine. An impression of the loss of

independence or freedom to have life once he starts using NIV was depicted. This image of reliance on NIV was also captured by Catherine, referring to NIV as something to be relied on for 'the rest of your days'. Furthermore, the image of NIV was strongly related to the end-of-life stage:

I remember his, um, he was really breathless almost all of the time in the end, towards the end, and fighting, you know, really fighting to breathe sometimes actually – Hanging onto life really I suppose. You know, despite all how he was the rest of him. That surprised me actually. So, what I thought was that I didn't want to end up like that gentleman in the home, fighting for breath and clinging on to life like that. He was in my mind a bit. (Catherine, pre-trial)

She expressed her surprise at the reaction of the patient with MND who was desperate to live, indicating her differing preference if she were in his position. Rather than prolongation of life, her concern seemed to be the quality of life. For Catherine, life is not worth fighting for when breathing is all that is left to her. Indeed, it was verbalized by Catherine that 'acceptability' of status and 'dignity' were important aspects in her dying process and NIV was negatively perceived in this context.

Personal perception of intervention with NIV: '...this is making me worse not better'

Some of the common reasons ($n = 7$) for withdrawal from NIV were the result of participants' perceptions of NIV treatment. Most participants expressed concern with some features of NIV, such as the mask, yet with variance in the level of their concerns. For Simon, this was addressed only briefly, indicating this to be a subordinate negative aspect of NIV to him. Likewise, Catherine briefly mentioned her preconception of the mask:

It's scary having something like that on your face I think. And I'd imagine it's quite claustrophobic as well; yeah, just having to wear that thing – doesn't sound very nice to me. (Catherine, pre-trial)

Catherine raised a potential issue of the claustrophobic nature of the mask. This was also an issue for Ted, as he elusively explained:

Ted: It's just something I just. . . it might be something I am frightened of. I don't know what it is – as though you trap something.

Interviewer: You trap something?

Ted: You know some people trap when they go into dark place or something like that. People. . . what's they – claustrophobia – I just don't want it. I don't want it on my face. (Ted, post-trial)

There was no clear explanation for his claustrophobia, and his experience was confused and emotional, as reflected in his poor articulation and the further observation that he could not pin down the exact phenomena he was concerned about. The illustration of being trapped, however, depicted the sense of insecurity and anxiety. As a result, he simply stated that he did not want it. This somewhat impulsive reaction towards the mask was also mentioned by Ian, who articulated the unbearable experience of having the mask on his face:

I told [doctor] at first when I saw it I said, “no way that I can stick the mask on my face”. I said “Foreign” “Alien” I just couldn’t do it. (Ian, post-trial)

He described the mask as ‘foreign’ and ‘alien’, expressing the impossibility of assimilation with the mask. Moreover, Ian later referred to the mask as ‘KGB torture’, implying the psychological and physical agony related to NIV. While the focus of these patients was on the negative features of the mask, Matthew described his concern as below:

I don’t want to ruin it and I don’t want to feel sick. I didn’t actually be sick, but it made me feel like I would be. So, I thought I wouldn’t use it – better not. I didn’t really like that. I thought “Well, this is making me worse not better” that’s what I thought you see. (Matthew, post-trial)

Even though Matthew did not actually vomit, his worry about being sick with the mask on was enough for him to decline NIV treatment. Moreover, this fear of being sick convinced him that NIV was actually making his health worse when he tried the machine at the trial. Because of this fear, he was reluctant to pursue NIV use regardless of the fact that the trial had taken place without him being sick.

The air pressure of NIV was also a cause of distress:

It was forced. . . . That’s what it’s like when it winds. It’s like, you know what I mean, I can’t breathe [Became upset]. (Rachel, post-trial)

I definitely don’t want that again. It’s just [pause] as if it stopped me breathing; as though I was trying to breathe out and this was blowing back and the wind was blowing out [exhale]. (Ted, post-trial)

Ted illustrated how asynchrony with NIV was experienced – as a fight to breathe. And considering the significance of breathing, the NIV machine was seen to invade the autonomy of his existence, and thus, he reacted against it. Based on this experience, Ted showed no interest in trying NIV again regardless of his knowledge that NIV would synchronize with his breathing pattern with acclimatization. This observation regarding the fear of losing autonomy over breathing can be also applied to Rachel.

Negative experiences with the health care service: ‘they all forced on me. . .’

Six participants revealed their dilemma over working with the hospitals. Both John and Peter reported receiving poor hospital service, and this was so influential that neither of them would even consider a trial of NIV. Moreover, they later withdrew from continuing with the study. This reluctance was already observable with John in his withdrawal from the formal interview process and the field notes describe this:

When asked about breathing tests he was reluctant to talk about them. He confirmed that this was related to his experiences at the hospital when prompted. He said that this had not been a very enjoyable experience for him and had put him off future visits. (John, pre-trial)

Likewise, Peter shared his encounter with the hospital:

I was agreeable [to have breathing assessments] at first but every appointment since 2006 has been cancelled. (Peter, pre-trial)

Peter reported to have been 'let down', 'disappointed', and 'upset' by the repeated cancellations, which consequently dampened his initial willingness to attend the assessments in consideration of NIV initiation. His response of rejecting NIV was a response to what he perceived as a previous rejection of his needs by the hospital. As a result, Peter actively chose to disengage with the health care service altogether to prevent further disappointment and the potential psychological distress this engendered. A similar explanation may be applied to John's behaviour, but for Ian, it was the image related to hospitals that affected his sense of security that discouraged him from going through the trial:

I just really have never liked them [hospitals]. They are places where you go when bad things happen really, aren't they? You know, when you're not well and people are dying and things. So, I mean I just don't like them. Never have. (Ian, pre-trial)

He showed his dislike of the idea of going into hospitals as an inpatient for a trial, for that would imply that he was becoming one of the dying people. Despite having no actual negative experience of hospitals, he reported that he had a 'phobia' of hospitals, which appeared to have originated from an existential concern. Hence, Ian avoided hospitals to protect his sense of mortality. Ian also described some negative interactions with medical staff as a negative aspect of hospital service. He perceived a violation of his autonomy and expressed frustration over the persistent approach of the hospital regarding NIV:

They're trying to persuade me, yeah, very much so. I know they're all at it, trying get me to change my mind – even though really I think it's my decision. And I have tried to tell them. I do say the same thing every time I go there. (Ian, pre-trial)

Ian spoke about the persistent and forceful approach from the hospital regarding potential NIV usage, and it was seen as an assault on his rights to make his own decision. The hospital was preventing him from living his life as he desired; thus, the health care service was seen to challenge his autonomy. A similar impression of intrusion into autonomy was shared by Simon in terms of hospital appointments made for him:

I'm tired of it all already. And it's going to get worse – I don't need all of it. I wish they'd leave me alone. If I haven't [long pause] if I haven't got long, I don't want to spend my time like that. (Simon, pre-trial)

The sense of him having an overwhelming number of appointments was apparent. It was interfering with his quality of life, and also it was creating a sense of loss of his autonomy. More importantly, however, it was likely that his awareness of the ultimate outcome of MND, being reflected by his silence, was the main reason for his desire to be left alone for his limited time.

Not Needing NIV: 'I don't think I need it'

The majority of the patients ($n = 7$) were dubious about the need for NIV. Among them, however, different reasons were identified for dismissing the idea. John witnessed no change in his breathing.

When questioned about his breathing, he felt that he had not experienced any change in his breathing and was not having any problems at all. When questioned about the previous symptoms at night mentioned by his wife, John denied knowledge of these. He felt he had not had any problems. Although it was perhaps natural for him not to consider NIV as he had no awareness of his breathing changes, his denial of his wife's account in the interview may indicate his refutation of the symptoms. Even if he was indeed not aware of the event, he insisted that there was no change in his breathing rather than taking notice of his caregiver's observation. Catherine experienced poor breathing, yet she still did not perceive a need for NIV as it was not problematic:

Actually my own breathing is quite good; I do get slightly breathless if I have to do too much, and if I get upset, but actually it doesn't cause me any problems or any worry or any kind of, um, it doesn't upset me. I mean, I'm usually just sat here during the day or I'm in bed and it doesn't cause me any problems here or in bed, I'm quite comfortable in bed. I mean, the reason I think they wanted to check my breathing was because I was having this little bit of breathlessness. But since [doctor] has started me on these anti-depressants, actually it's almost completely gone. (Catherine, pre-trial)

Although Catherine acknowledged she had episodes of breathlessness, she showed little concern about it and stated that it was actually 'quite good'; her breathing was not causing her problems. Simon similarly admitted a change in his breathing, yet did not perceive a need for support from NIV treatment:

I do occasionally [get short of breath]. But I have a spray for that, to help me if I feel my breathing is slightly short. I have a spray for that. So, because I can use that, I don't have any concerns at all about my breathing really. (Simon, pre-trial)

Simon occasionally experienced more than mild breathlessness that needed addressing; yet a spray originally prescribed for his asthma effectively cancelled the impact of breathlessness; thus, he reported no concern over his breathlessness: his breathing was recoverable and manageable. Other patients also shared their doubts:

I'll try anything if it will help me. But the thing is that some days I'm alright getting out of the chair and some days I struggle a bit. So I think "Do I really need a chair?" It's like that a bit with the breathing as well. Some days I feel better than others, so I think "Well, do I really need a machine?" Because they did these overnight tests, but what if they were just one of my bad days, and the rest of the time I could be alright, couldn't I? So I don't know about all these new things coming to me. (Matthew, post-trial)

Even though Matthew showed his willingness to employ aids, he questioned their necessity. He implied a conditional acceptance of the use of aids, indicating his resistance. Also, his definition of good days and bad days, based on his experience of the days, is seen to be superior to the overnight test presumably because of the daily assessment compared with the one off assessment. This doubt regarding the credibility of hospital assessment was held by other patients, with one patient even carrying out his own assessment for confirmation:

Well, it's [breathing] pretty stable. You know, it's stable on that thing [peak flow meter]. I've taken a few this week and it's been stable. (Ian, pre-trial)

After being recommended to use NIV, Ian started his own breathing assessment with a peak flow meter and the stable results gave him confidence in his breathing. For Ian, his own measurements, taken on more than one occasion, confirmed that his breathing was fine. In contrast to these patients whose arguments were based on their understanding of their condition, Henry declined NIV after a medical consultation. In addition, Henry appreciated the benefits of NIV and used it for some time until his condition dramatically changed:

Now they've [doctors] told me that I don't have to use it [NIV]. (Henry, post-trial)

In consideration of the progressive weakness outweighing the benefits of NIV, Henry declined the treatment as it had stopped being effective and the focus of his treatment was presumably changed from management of symptoms to the management of dying.

Overall hermeneutic circle

Although each superordinate theme derives from a different element of patient experience leading to a rejection of NIV, a key driver was the maintenance of the self, that is, the maintenance of autonomy, dignity, and quality of life. Firstly, the sense of self was affected by the illness itself through the sense of losing control over the self due to the illness. Consequently, patients became reluctant to engage with NIV, a treatment identified as a further threat to self. Secondly, some features of NIV were perceived to generate insecurity and impair quality of life. Needing the machine and mask interface was perceived to challenge their autonomy. Furthermore, unpleasant interactions with health care services were perceived by some participants to be assaults against the self, causing emotional distress and disempowerment to patients and, as a result, creating a general desire in the participants to avoid interaction with health care. Finally, the perception that NIV was not necessary can be interpreted as an attempt to preserve identity and independence by rejecting the idea of needing NIV.

Discussion

The aim of this study was to understand why some patients decline NIV, a potentially life-extending treatment, when it is clinically indicated. IPA identified four themes: preservation of the self, negative perceptions of NIV, negative experiences with health care services, and not needing NIV. In this study, the term 'self' focused on autonomy, dignity, and quality of life, which were identified to be influential in how patients evaluate their experience of using NIV. As mentioned earlier, we accept that the interpretation presented here is not the only interpretation of the accounts and that others, including those that interpret refusal of NIV as being due to the way it raises the threat of mortality, may be possible.

Based on the model introduced by Gauthier and Swigart (2003), it is argued that the participants in the current study were reluctant to engage with the treatment because they were reluctant to admit their own condition and its consequences (*realization of terminality*), and they were unwilling to adapt to the changes and accept the need for help (*accommodating living*). The need for NIV was dismissed mostly because they did not perceive their condition as problematic, if at all. It is interesting to note that most participants acknowledged changes and to some extent accepted them, yet showed a

reluctance to acknowledge them as a symptom of MND. It may be argued that this tendency was related to their attempt to maintain the self, which had been damaged by MND, rather than preparing for the unknown future (*engagement with uncertainty*). Both control over the treatment and quality of life were also observed as important aspects in decision-making process regarding NIV usage, and these are elaborated upon below.

The frustration over their new identity associated with their impaired independence, as well as the fear of becoming dependent on NIV, was explicitly displayed by participants. The health psychology literature provides evidence that disease can cause a dramatic change not only biologically, but also psychologically (Dickson, Allan, & O'Carroll, 2008; Harman & Clare, 2006; Williams, Moffatt, & Franks, 2004). An emotional reaction to loss of independence and its impact on the way the self is perceived is seen in various illnesses (Dickson *et al.*, 2008). A common phenomenon after diagnosis is appraisal of the situation through comparison with previous self or others (Harman & Clare, 2006; Williams *et al.*, 2004). Such a comparison may direct patients to preserve those aspects of their self that disease cannot readily corrupt: their autonomy, dignity, and some aspects of quality of life. Preservation of the self, in these terms, was clearly very important to these patients and was a large part of declining and withdrawing from NIV.

Anxiety that NIV would be a threat was influential in decision-making. A significant reason for withdrawal was inherent to the treatment itself (i.e., the mask and the forced air pressure), and this was not simply related to comfort issues. Rather, their negative responses to NIV were generated as a result of fear and a reflection of their threatened autonomy. The dominant negative feature of the mask expressed in the current study has been previously reported as a cause of poor adherence to ventilation (Chasens, Pack, Malislin, Dringes, & Weaver, 2005). Furthermore, irrational behaviour and a feeling of panic with the NIV mask has been argued to be the manifestation of anxiety and insecurity associated with the feeling of being powerless and vulnerable in a given situation (Torheim & Gjengedal, 2010). In line with this, high adherence to ventilation was found to be related to individuals with higher self-efficacy (Wild, Engleman, Douglas, & Espie, 2004).

Poor impression of and interaction with health care services were found to challenge the sense of self by causing emotional distress and disempowerment in the forms of invading autonomy and the loss of control. Negative experiences with health care services were described in association with withdrawal from NIV, and indeed treatment of MND. Participants described the overwhelming nature of appointments and persistent persuasion to fall in line with the prescribed treatment, each serving to challenge self-value. Previous negative experiences were also perceived to have contributed to their decision to decline further involvement with health care services. 'Devaluation' generated by health care services causes dissatisfaction, identity threat, and disempowerment in patients (Coyle, 1999), which ultimately was addressed by patients with withdrawal from services leading to disengagement with formal health care.

The need for NIV was doubted by all but one patient. Some patients did not feel as though they had sufficient symptoms to warrant a need for treatment. It is possible that their breathlessness was covert due to a gradual adaptation to their respiratory changes as found with chronic obstructive pulmonary disease (Gysels, 2008). Alternatively, some individuals may not see the need for NIV due to their confidence in their ability to improve the problem (Prins, Verhaak, Bensing, & Meer, 2008). An attribution to a different causation, such as finding an alternative explanation for breathlessness, is often observed when people try to make sense of the experiences they are facing (Gysels, 2008; Horrocks, Somerset, Stoddart, & Peters, 2004). Critically, this perception was linked with declining or withdrawal from NIV.

The important role the self played in the decision-making process in this particular group was acknowledged. The detail of their negative responses to NIV differed, yet all the accounts demonstrated that what mattered in their decision-making stemmed from the same concern: maintenance of the self. This general finding reaffirms that an idiographic approach reflected by hermeneutic practice was crucial in understanding these particular individuals in the particular setting of decision-making regarding NIV in the context of MND.

Limitations of the study

The employment of the in-depth interviews enabled the researchers to understand plausible reasons for NIV withdrawal, yet the limited number of post-NIV trial accounts necessitates some caution. Although available post-NIV data from other patients showed overall agreement with this analysis, additional themes may emerge from more post-NIV interviews. Also, almost all of the data reported in this study were collected from patients who had declined or withdrawn from NIV treatment at an early stage. Further studies should include patients, who decline the treatment, from different stages of NIV use to assess whether the reasons identified in the current study are still applicable. Studies exploring how the concept of both identity and dependency issues are being dealt with by NIV users are also warranted.

Another limitation of the study is restricted verbal communication of some patients, including John, who declined from a formal audio recorded interview. Inclusion of all the participants who declined the treatment was essential for the purpose of the study, yet the rich information we were looking for was not available on account of this issue.

Conclusion

Just under a third of patients in the cohort we have followed did not wish to make use of NIV. Given the proven survival benefit from NIV, this was an unexpected observation, yet not unique to this study. The main issues with NIV use were recognized to be how their sense of self was perceived to be affected by the use, or potential use, of NIV. Furthermore, preservation of the self was not only perceived to be relevant in terms of NIV use, but also in how they understood the phenomena of living with MND. The issues raised about previous experience with health care services are salutatory for those involved with such patients.

Based on the findings, we suggest that it is crucial to understand the psychological dimension involved in decision-making regarding the use of NIV and would suggest a sensitive holistic evaluation if NIV is declined. In addition, efforts could be made by manufacturers of NIV equipment to take the tendency for claustrophobia into account by finding designs that mitigate this reaction. Furthermore, it may even be possible for designers to find a way of supporting identity despite the need to use the equipment. Further work to determine whether supporting their sense of self would alter patients' decision-making on NIV would be beneficial as services seek to improve their care to individuals with MND.

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