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## Exploring and addressing 'concerns' for significant others to extend the understanding of quality of life with motor neurone disease: a qualitative study

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Complete List of Authors:	Ando, Hikari; University Hospital Aintree, Respiratory Laboratory Cousins, Rosanna; Liverpool Hope University Faculty of Science, Psychology Young, Carolyn; The Walton Centre NHS Foundation Trust
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Abstract:	<p>Background: The absence of curative medication for amyotrophic lateral sclerosis (ALS) makes palliative care and understanding quality of life (QoL) in ALS a clinical priority. Previous qualitative research has explored the concept of QoL in terms of illness impact on life perspectives and sense of self. Objective: In this research we explored 'concerns' – one of the four aspects in the World Health Organization's conceptualisation of QoL – towards adding to the literature. Methods: In-depth interviews with 26 individuals with ALS were subjected to thematic analysis involving both inductive and deductive approaches to explore participant's concerns, and to evaluate the relevance of their concerns for understanding QoL in ALS. Findings: The analysis showed that concerns for significant others contribute to participant's QoL because of their existential value. It was important for participants to minimise the impact of limitations and burdens associated with ALS on significant others, even at a cost to self. Discussion: The current study supports a holistic approach in service provision, ensuring the inclusion of relevant significant others. It is further suggested that clinicians explore the specifics of burdens perceived by patients in order to support them in minimising the burdens for their significant others.</p>

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## Abstract

**Background:** The absence of curative medication for amyotrophic lateral sclerosis (ALS) makes palliative care and understanding quality of life (QoL) in ALS a clinical priority.

Previous qualitative research has explored the concept of QoL in terms of illness impact on life perspectives and sense of self. **Objective:** In this research we explored ‘concerns’ – one

of the four aspects in the World Health Organization’s conceptualisation of QoL – towards adding to the literature. **Methods:** In-depth interviews with 26 individuals with ALS were subjected to thematic analysis involving both inductive and deductive approaches to explore participant’s concerns, and to evaluate the relevance of their concerns for understanding QoL in ALS. **Findings:** The analysis showed that concerns for significant others contribute to participant’s QoL because of their existential value. It was important for participants to minimise the impact of limitations and burdens associated with ALS on significant others, even at a cost to self. **Discussion:** The current study supports a holistic approach in service provision, ensuring the inclusion of relevant significant others. It is further suggested that clinicians explore the specifics of burdens perceived by patients in order to support them in minimising the burdens for their significant others.

## Background

In the past 10 years, an enormous amount of research has been dedicated to understanding amyotrophic lateral sclerosis (ALS), also known as motor neuron disease (MND), a life-limiting neurodegenerative disease underpinned by the loss of motor neurons. However, to date, there is neither cure nor effective disease modifying therapy. Prevalence rates for ALS are estimated to range from 2.6 to 3 cases per 100,000 people<sup>1</sup>. Incidence increases with age,

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3 with a peak at 47-63 years, and new diagnosis after 80 years of age is rare<sup>2</sup>. Regarding  
4 survival rates, about half of those diagnosed with ALS die within 30 months, although a fifth  
5 will live beyond 5 years after the symptom onset<sup>3</sup>. There are three onset types in ALS which  
6 leads to variety in type of support requirements, at least initially. These are limb onset (about  
7 70%), bulbar onset (about 25%), and respiratory onset (5%)<sup>2</sup>. Those with limb onset may  
8 experience difficulty in walking, foot drop, or loss of fine hand movements. Individuals with  
9 bulbar onset usually display dysarthria or dysphonia. Dysphagia often accompanies  
10 dysarthria and is typically more pronounced with liquids than solids. Respiratory onset is the  
11 least common mode of onset, involving breathlessness and nocturnal hypoventilation, which  
12 may be associated with hypersomnolence and fatigue. In addition, for some patients there are  
13 cognitive symptoms which have been linked to a shared aetiology of ALS and frontotemporal  
14 dementia<sup>4,5,6,7</sup>. Prolonged survival is not restricted to individuals who have a milder disease  
15 progression; some long-term survivors are extremely disabled with progressive paralysis,  
16 bulbar weakness and respiratory failure, but life is sustained through use of parenteral feeding  
17 tubes and ventilatory support.

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39 A diagnosis of ALS is devastating, and the negative impact extends beyond the  
40 individual to their family and close friends<sup>8</sup>. In the clinic, the focus is symptom management  
41 and the preservation of quality of life (QoL) for the patient and their family caregiver. The  
42 importance of QoL is signified in UK National Institute for Health and Care Excellence  
43 (NICE) guidance<sup>9</sup>; it suggests the concept is used as an indicator of effective treatment<sup>10</sup>.  
44 However, the construct of QoL is yet to be established, and therefore the use of  
45 measurements without an established structure of QoL leads to the argument that QoL is  
46 defined by the measurements employed for the study<sup>11</sup>. Despite a lack of consensus on the  
47 conceptualisation of QoL, researchers agree that it is a multidimensional construct and QoL is  
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3 generally understood to be a subjective evaluation of one's own life<sup>12,13,14,15</sup>. Objective  
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5 aspects of one's life are evaluated using cognitive appraisals, and these appraisals use  
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7 affective and cognitive components in the evaluation process<sup>12,16,17,18</sup>. Based on the  
8  
9 understanding that the concept of QoL is subjective, some research on QoL in ALS utilised  
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11 quantitative methodologies which have used instruments such as the Schedule for the  
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13 Evaluation of Individual Quality of Life (SEIQoL)<sup>19</sup> to discern predictors of QoL, arguing  
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15 that this allows researchers to employ an idiographic approach<sup>20,21,22</sup>. Nevertheless, further  
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17 exploration is necessary if one seeks to fully understand the underlying significance and  
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19 rationale for the factors proposed to improve QoL in NICE guidance.  
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24 Besides taking a quantitative approach, the literature includes qualitative studies  
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26 which provide an understanding of the subjective evaluation of life circumstances with ALS.  
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28 The focus has generally been on the physiological impact of living with ALS on life  
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30 perspectives<sup>23,24,25</sup> and perceived sense of self<sup>26,27,28</sup>. Previous qualitative research on life  
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32 perspectives in ALS has centred on its fatality<sup>23,24,25</sup>. Knowledge of their prognosis has been  
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34 found to provoke distress amongst patients<sup>25, 28,29</sup>, with some individuals experiencing a  
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36 further challenge to their existential meaning<sup>25</sup>. In addition to the threatening picture of their  
37  
38 future, the impact of the ALS illness trajectory on how people perceive themselves has been  
39  
40 explored. A challenge to previous standards of self is a frequently experienced phenomenon  
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42 in which the current situation is evaluated against 'normal' life before the illness<sup>28</sup>. Losing  
43  
44 the ability to perform tasks in the way they had been done previously often leads individuals  
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46 to perceive their identity to be eroding away with ALS<sup>26,27</sup>. This identity threat can provoke a  
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48 further challenge to their existential meaning of some individuals with ALS<sup>23,26</sup>. Interestingly,  
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50 previous research suggests a sense of being a burden to others because of their need for  
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52 physical support can have an influence on how individuals evaluate their QoL<sup>25,30,31</sup>;  
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3 nevertheless, individuals were able to find meaning and self-value if they were able to make  
4 contributions to others<sup>25</sup>. Therefore, sense of self can be violated, reconciled, or even  
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6 enhanced depending on the nature of interactions. Similarly, existential meaning may be  
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8 either established or threatened through inter-personal relationships.  
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13 Findings from previous qualitative research indicate that the quest of QoL research in  
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15 ALS is to explore whether the condition challenges people's previously held beliefs about the  
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17 meaning of life. And, if changes associated with ALS lead to a reconstruction of subjective  
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19 meanings of the world, then how does this impact upon the evaluation of one's life quality.  
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21 The constructivist view asserts that people actively construct realities through revisions of  
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23 their understanding of the world when their previous understanding is proved to be  
24  
25 insufficient<sup>32,33</sup>. There are multiple realities that are subjective to an individual<sup>34,35</sup>, although  
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27 subjective constructs do not provide objective reality - simply that "all constructed meanings  
28  
29 reflect a point of view" (p. 2)<sup>32</sup>. Therefore, QoL is understood to be the subjective meaning of  
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31 life against which one's own life is evaluated. This constructivist understanding of QoL  
32  
33 echoes the process of evaluation of QoL described by the World Health Organization in  
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35 1996<sup>36</sup>. QoL is defined as "an individual's perception of their position in life in the context of  
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37 the culture and value systems in which they live and in relation to their goals, expectations,  
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39 standards, and concerns" (p354).  
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46 Referring to the WHOQOL definition, it is argued that previous qualitative research  
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48 has extensively explored the contribution of 'expectations' and 'standards' to QoL in ALS  
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50 under the concept of life perspectives and the illness impact on perceived sense of self,  
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52 respectively. There is, however, relatively little research on 'goals' and 'concerns' – the other  
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54 two aspects of QoL<sup>36</sup>. In particular, little is known about the significance of 'concerns'  
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56 although the aspect has been touched on by previous qualitative research<sup>37,38</sup>, as cited in the  
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3 qualitative thematic synthesis by Soundy and Condon<sup>39</sup>. However, these results need further  
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5 exploration to understand ‘why’ and ‘how’ of the significance of ‘concerns’ to understanding  
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7 QoL in ALS. To date, the most informative study on this aspect is found in a study of burden  
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9 in ALS by Foley and her colleagues<sup>31</sup>. Interviews with 34 individuals with ALS illustrated  
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11 how participants with ALS were aware of stress in family members, and many provided  
12  
13 emotional support to alleviate their distress to an extent that the preference of the person with  
14  
15 ALS was knowingly compromised. The aspect of end-of-life care and its impact on their  
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17 family were observed to generate anxiety among participants – especially where they had  
18  
19 young children. The authors argued that a sense of duty as a family member underpinned  
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21 manifest contributions and decisions, and this reflected the significance of the social context  
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23 in which their participant’s self and self-value was influenced. This suggests a close link  
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25 between ‘concerns’ for significant others and ‘standards’ for sense of self. Models of  
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27 caregiving<sup>40</sup> illustrate that quality of life outcomes for patient-caregiver dyads are mutually  
28  
29 dependent. This strongly suggests reciprocity in the ALS illness situation, and that  
30  
31 understanding the role of concerns for significant others is an important part of understanding  
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33 QoL for the ALS patient, and ultimately providing timely support. A further exploration is  
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35 needed to better understand whether ‘concerns’ is a separate important aspect of QoL, or if  
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37 this can be indeed merged into the aspect of ‘standards’ in lives of people with ALS.  
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45 To summarise, it is understood that people engage with meaning-making activities to  
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47 construct and reconstruct realities of the world, and this subjective meaning-making process  
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49 influences how they evaluate their lives (i.e. QoL). Previous research has informed our  
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51 understanding of how ALS may affect people’s evaluation of their lives because of altered  
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53 expectations (i.e. life perspectives) and standards (i.e. sense of self). In order to gain more  
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3 informed knowledge on QoL in ALS, the present manuscript was prepared to qualitatively  
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5 explore how the WHOQOL aspect of ‘concerns’ is relevant in the lives of people with ALS.  
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## 8 9 **Method**

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12 This study was conducted at a National Health Service (NHS) Foundation Trust covering  
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14 North West England, UK. It contributes to a longitudinal mixed-methods investigation of  
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16 QoL in people with various neurological conditions, including ALS. The full study uses an  
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18 exploratory sequential approach<sup>41</sup>; the qualitative phase was conducted first to explore factors  
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20 that are influential to QoL in people with ALS. This was deemed as appropriate given that  
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22 the concept of QoL is subjective.  
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26 In this manuscript, one of the four themes identified in the main study is presented towards  
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28 extending current understanding of the importance of this aspect in ALS. This theme -  
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30 concerns for significant others - aligns with the WHO definition of QoL aspect of ‘concerns’.  
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32 The other three themes – perceived illness prognosis, sense of self, and life to enjoy – will be  
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34 presented in a separate paper explaining the overall findings of the qualitative phase of this  
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36 study.  
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## 41 **Ethical approval**

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43 The study was approved by the National Research Ethics Service (NRES) for the North West  
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45 region of the UK (NRES reference number: 11/NW/0743). Potential participants were given  
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47 an information sheet providing full details of the study. Confidentiality and anonymity were  
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49 assured, as well as a clear explanation of right to withdraw, in advance of asking for written  
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51 consent.  
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## Sampling

Inclusion criteria were a diagnosis of either bulbar or limb onset ALS in any stage of the disease, and an ability to communicate with a researcher. Due to the low prevalence of respiratory onset ALS, no participant with the respiratory onset type was recruited to the study. Verbal communication could be supported by writing or the assistance of a caregiver. Individuals were excluded if they were not capable of informed consent, or they were suffering from a concomitant serious medical or psychiatric condition.

## Recruitment setting and procedure

Recruitment was at the outpatient clinics at the study site, at the time of a scheduled appointment with a neurologist. Following their consultation, the study was introduced by the neurologist, and potential participants who showed an interest in the study were invited to meet with a researcher for further information about the methodology. Purposive sampling was included to ensure appropriate consideration of onset types to ensure that a wide diversity of ALS experience was included. Based on the argument that 12 interviews are sufficient for data saturation for thematic analysis<sup>42,43</sup>, the current study aimed to recruit at least 12 participants for both bulbar and limb onset types.

## Data collection

In-depth semi-structured interviews were used to understand and explore factors that are important for QoL of people with ALS. An interview schedule (Appendix 1) was developed by two psychologists based on the literature review that QoL is a subjective evaluation of one's whole life involving various aspects. As such, we used open-ended questions that permitted discussion of any and all aspects of participant's quality of life. There was no attempt to define quality of life, this was solely their interpretation with encouragement to



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2  
3 elaborate when appropriate. Given that accessible information gained through interactions  
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5 with participants depends on their willingness of revelation and that some information could  
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7 remain hidden<sup>33,44</sup>, seven of the 13 person-centred interaction styles<sup>45</sup> were used to build a  
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9 research-participant relationship. These interaction styles were affirmation attention,  
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11 checking understanding, restating, providing reassurance, acknowledging participants'  
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13 unstated feelings, direct questioning, and maintaining silence. The interviews lasted 20 – 90  
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15 minutes; individuals who used communication alternatives took longer. Face-to-face  
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17 interviews at the study site were preferred, however, participant's preference was respected,  
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19 and eight interviews were conducted over the telephone to accommodate their wish not to  
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21 travel to the study site.  
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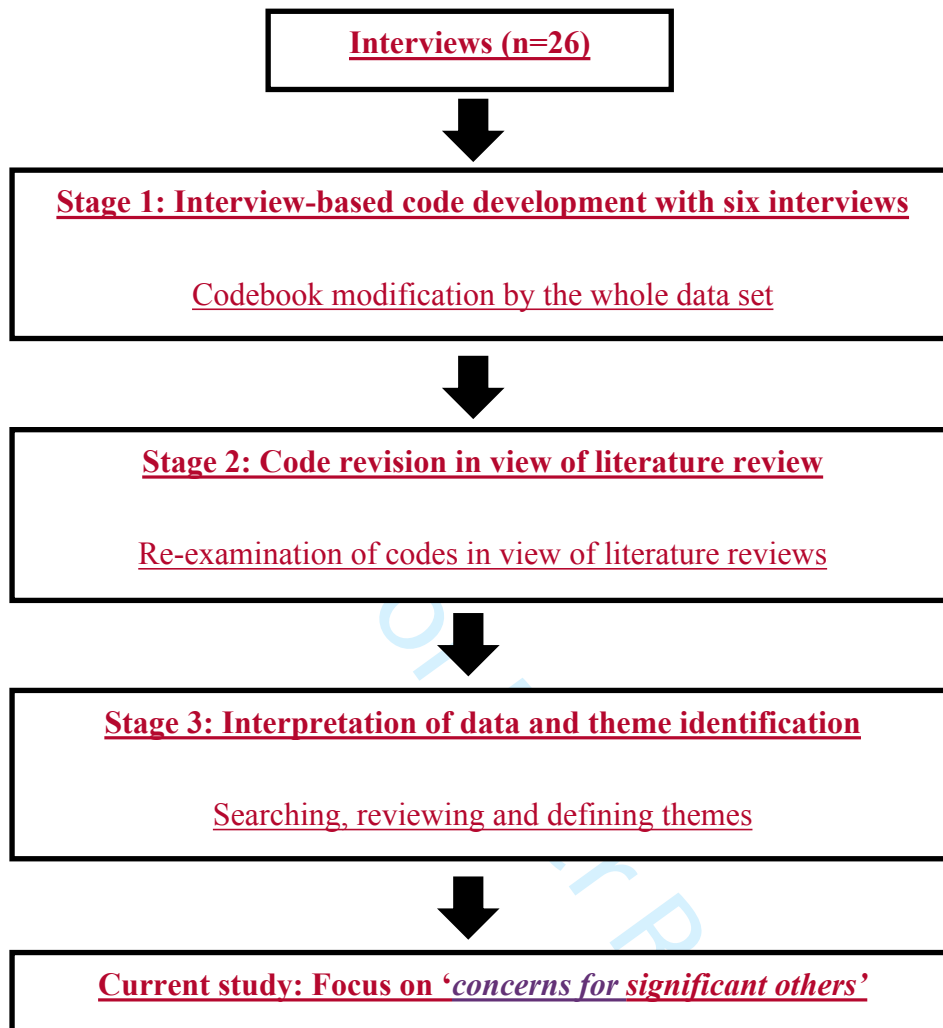
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27 Efforts to mitigate communication difficulties were made, especially with bulbar  
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29 onset ALS patients. The study embraced the argument of employing alternative  
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31 communication modes to capture experiences of individuals with varying symptoms<sup>46</sup>. The  
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33 alternative communication modes employed were: extra active listening, allowing more time  
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35 for the participants, communication alternatives (i.e. writing and text-to-speech devices), and  
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37 accepting input from companions. Five participants employed either writing or a text-to-  
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39 speech device to support their communication. Most participants were interviewed alone; six  
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41 participants attended the interview with a companion. The decision to respect the  
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43 participants' preference to have the third person present during the interview was based on  
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45 ethical considerations of the need of physical and emotional support for the participants,  
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47 reflecting the notion of 'vulnerable' participants in a health research setting<sup>47</sup>. Despite the  
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49 benefits of support from a companion, especially where they were able to help the  
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51 interviewer understand the interviewee, it was recognised that the presence of the companion  
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53 was a potential hindrance to an open conversation<sup>48</sup>. In line with this, the presence of a  
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3 companion is noted in the finding section in consideration of their potential influence on a  
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5 participant's narrative.  
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8 **To fully understand the context of ability and disability in the ALS illness situation**  
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10 **we also administered the revised ALS Functional Rating Scale (ALSFRS-R) to all consenting**  
11 **participants. This scale measures physical functioning of daily activities<sup>49,50</sup>; the scale ranges**  
12 **from 0 to 48 with higher scores indicating better physical functioning.**  
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## 18 **Data analysis**

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21 This study employed a modified thematic analysis<sup>43,51</sup> to determine what is important  
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23 for the QoL of people with ALS. The analysis incorporated inductive and deductive  
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25 processes<sup>52</sup> involving two full analyses of the same data to form an understanding of QoL for  
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27 a more informed consensus (See figure 1). This approach reflects the current research  
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29 paradigm of constructivism. During the analyses, the primacy of the inductive approach was  
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31 perceived given that the concept of QoL is subjective, and therefore the analysis began with a  
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33 bottom-up approach. The inductive approach was reflected in the bottom-up development of  
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35 a codebook which permitted a systematic analysis of all the information collected. In our  
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37 analysis, codes from the first six interviews formed the baseline codes for the codebook  
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39 instead of reviews and evaluations of themes as outlined by Braun and Clarke<sup>51</sup>. It has been  
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41 argued<sup>43</sup> that data from six interviews is sufficient to support theme development, with  
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43 additional interviews then assisting the refinement of the findings.  
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**Figure 1: Flow chart of the analysis procedure**

The baseline codes from the six interviews were applied to the remaining interviews and modifications to codebook were made as required. Extracts for codes were reviewed in order to examine whether each code was coherent, yet distinct from others. Following the inductive analysis, the codebook was reviewed and modified in view of previous research including WHOQOL's definition of QoL for a systematic examination and identification of the influential factors of QoL. The modified codes were applied to the whole data set using NVivo for Mac (version 11.2.1). Once the codes were finalised and applied to the whole data set, the extracts for each code were examined. At this stage, identification of influential factors for QoL was undertaken by exploring links between the codes, using NVivo. All the

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3 quotes containing codes on QoL were retrieved and their links with other codes were  
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5 examined through reviewing extracts. Four overarching themes were identified from this  
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7 analysis. These were *perceived illness prognosis*, *sense of self*, *life to enjoy* and *concerns for*  
8 *significant others*. For this current article, codes for *concerns for significant others* were  
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10 retrieved and explored to understand how they were significant for QoL.  
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### 15 **Trustworthiness of the research**

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18 The constructivist's ontological claim of multiple realities, with the viewpoints of both  
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20 epistemology and axiology, implies that a researchers' view needs no justification of its  
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22 validity. However, the methodological position of a hermeneutic and dialectical approach  
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24 calls for a careful examination to assure the qualities of an interpretation through which a  
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26 reconstruction is made. It has been argued that researchers should ask themselves whether the  
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28 implications of their findings are good enough to pursue behaving in a different way<sup>53</sup>. In  
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30 other words, the trustworthiness of research should always be addressed. In view of  
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32 constructivism as the worldview of the current study, trustworthiness was sought through  
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34 focusing on **sampling adequacy**, researcher-participant relationships, reflexivity, thick  
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36 description and peer examination of analysis.  
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42 **Sampling adequacy is concerned with data saturation to ensure the identification of**  
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44 **common attributes for informed understanding of meaning-making process. In the current**  
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46 **study, this meant recruiting an adequate sample size to understand how the aspect of**  
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48 **'concerns' is relevant for QoL of people with ALS. The sample size for the current study was**  
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50 **determined based on previous reports<sup>42,43</sup>; data saturation was subsequently observed.** The  
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52 significance of the researcher's relationships with participants was considered given that  
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54 multiple realities are examined through researcher-participant dialogues. In particular, a  
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56 reciprocal relationship<sup>54,55</sup>, not a hierarchical relationship was sought towards establishing  
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trust in the relationship of research with participants. The employment of reflexivity reflects arguments that researchers enter a study with their own values to understand the data<sup>35,41</sup>, and thick description enables readers to evaluate the analysis of the data<sup>56</sup>. Although the rigor of analysis was scrutinised further through peer examination of analysis<sup>57</sup>, the constructivist position suggests that the findings will always be open to new interpretations to reflect multiple realities<sup>34</sup>.

## Findings

Thirty-five individuals with ALS asked for information about the study. Of these 28 consented and were interviewed. Two interviews were removed from the analysis due to failure of audio recording, giving a total of 26 participants. See Table 1 for the characteristics of the participants. The mean illness duration for the whole cohort was 1 year 8 months.

**Table 1: Demographics and baseline illness characteristics**

<b>Demographics</b>		
Male (%)	14 (53%)	
Mean age (range)	64yrs (39-84yrs)	
Illness onset		
Limb	13	
Bulbar	13	
<b>Illness characteristic</b>	<b>Median (IQR)</b>	<b>Range</b>
Illness duration	9.5m (24)	1m – 9yrs 2m
ALSFRS-R	38 (13)	12 – 48

*Note.* ALSFRS-R, the revised amyotrophic lateral sclerosis functional rating scale<sup>49,50</sup>

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3 The analysis found concern for significant others to be an important aspect of  
4 participants' QoL. Significant others were those with whom participants held a close  
5 relationship, and these are usually their partner/spouse, child(ren), parent(s), and  
6 grandchild(ren). Participants expressed the positive impact of having significant others in  
7 their lives, and they were concerned about the impact of the diagnosis on the well-being of  
8 significant others. The findings are presented below under four themes: the significance of  
9 significant others, greater importance of significant others' well-being, perceived illness  
10 impact on others, and minimising the burdens. The findings are elaborated upon with an  
11 indication of their meaning in terms of QoL and are illustrated by verbatim quotes from the  
12 narratives. Square brackets are used to either provide information for clarity or replace  
13 information to maintain confidentiality. The brackets with three full stops indicate an  
14 omission of words.

### 34 **The significance of significant others**

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37 The importance of having significant others was made manifest when people referred to their  
38 significant others as being what mattered most in their lives. For them, their quality of life  
39 depended on the presence of significant others. Many of our participants said some version of  
40 "As long as I have got my family – that will do me", and typically elaborated upon, as in this  
41 quote:  
42

43  
44 "I have a loving family and I see plenty of them, I see them most days, I see my  
45 daughter every day and I see family most days and they add to my quality of life [...]  
46  
47 As long as I've got my family that's all that matters to me."

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3 Whilst participants expressed the significance of important others in their lives, the following  
4 participant reflected on how this may be neglected prior to the diagnosis because of the  
5 current social climate where people are largely “work and materially driven”. He told us:  
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11 “Life prior to MND was so busy and oriented by what you are going to do in the  
12 future, trying to earn enough money for holidays and paying the mortgage. After  
13 diagnosis you realise that those things are not the be-all and end-all of things and that  
14 your relationship with your family is far more important.” (Accompanied)  
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21 Following a diagnosis of ALS, his life priorities were revised and he perceived his  
22 relationship with his significant others to be of most value. Elsewhere in the interview the  
23 participant indicated that this realisation was partly facilitated by the fact that he was  
24 spending the majority of his time with his significant others, mainly his partner, involving  
25 intimate and personal care. His interview therefore suggested that his special relationship  
26 with his partner was further nurtured through sharing more time, therefore their lives,  
27 together.  
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39 “I have a very good relationship [with partner] – I think that has become very much  
40 stronger since the disease was diagnosed in some ways. Prior to January last year, I  
41 was working 12-hour shifts, days and nights, quite often 5-6 days a week, [whereas]  
42 last 18 month now where we’ve been together probably 95% of the time.”  
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48 (Accompanied)  
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51 The importance of significant others and the sharing of life was further observed through  
52 interviews with two individuals whose significant others were absent: “I don’t have family  
53 here [where she lives]; I am on my own [upset]. Analysing [sic] it. My quality of life that I  
54 can’t do all the activities I used to enjoy; I can’t go home and see my mum” (accompanied).  
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3 Similarly, another participant who had lost her husband and son said: “Any sadness I fear I  
4 indulge perhaps once a week. Just have a little weep – not sorry for myself, just wishing that I  
5 could talk to, you know, my husband and my son.” These participants provided insight into  
6 how the absence of significant others created a hole in their lives which cannot be filled by  
7 anything else, highlighting the importance of significant others in people’s lives.  
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### 19 **Greater importance of significant others’ well-being**

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22 The importance of significant others was such that participants displayed their concerns for  
23 the well-being of their significant others, and this was observed in statements such as: “It  
24 upsets me to see her [wife] upset”. That is, the perceived well-being of significant others in  
25 turn, affected participant’s own emotional well-being. The quote indicates the close  
26 relationship between QoL of significant others and that of participants. This was further  
27 illustrated by another ALS patient: “She [wife] is everything to me. So I’m made up, I’m  
28 happy this is happening to me rather than to her”. The participant evidently appreciated the  
29 presence of his wife in his life; his wife was what he had and what mattered to him.  
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31 Furthermore, the quote suggests that his wife’s life was more precious to him than his own  
32 life given that he expressed a preference for his wife’s well-being over his own. This belief in  
33 the greater importance of others’ well-being was reflected in how participants talked about  
34 their general concerns for their significant others. One participant indicated his preference for  
35 living overseas, yet his concerns for his family kept him in the country. Another participant  
36 voiced his concern for his son with alcohol dependence, and when asked what his priorities  
37 were he said, “I’m having a particular problem with – I have a son who is an alcoholic and I  
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3 can't really deal with him now". For this participant, his son's alcohol dependence problem  
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5 was more concerning than his own illness.  
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### 8 **Perceived illness burdens on others**

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11 Concerns for significant others was common amongst participants. Their concerns were  
12  
13 related to both the present and the future. This two-levels of burdens is illustrated in the  
14  
15 following quote in which the participant described the altered previous normality as the  
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17 ongoing psychological burdens of ALS on his wife (present-oriented burdens), whilst also  
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19 noting the future-oriented dilemma (future-oriented burdens).  
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23 Interviewee: It's been harder for my wife to cope with [ALS] and I think if she was  
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25 ill it may be harder for me.  
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28 Interviewer: Why do you think that might be?  
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31 Interviewee: Well, we are very close. I think two things: partly she doesn't like to  
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33 see me weaker. For example, she remembers when my speech was  
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35 normal ... she forgets that she can't hear that normally. And I think she  
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37 misses that fact that I can't walk about too much ... There is also  
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39 uncertainty about how she will cope when I get worse.  
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43 Further explorations of each type of burden are expanded as follows.  
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46 **Present-oriented burdens.** Participants were particularly worried about illness-related  
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48 burdens on significant others and how these burdens may affect others' QoL. The burdens  
49  
50 were described in terms of restrictions, which were often created by workload due to the  
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52 physical demands of caring for someone with ALS. Participants acknowledged the amount of  
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54 work which fell on their significant others. Two examples:  
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3 “Obviously, that puts a bigger strain on my husband, who is doing massive [amounts]  
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5 so that worries me slightly.”  
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9 “I know I’ve got to live with it [ALS], but she’s [wife] got to do everything. She  
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11 never seems to have any time off.” (Accompanied)  
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15 As shown, these physical demands meant not only pressure on physical well-being, but also  
16  
17 implied an invasion into the day-to-day life of the significant others thereby restricting them  
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19 because of the additional demands. A potential impact of the illness on others was articulated  
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21 by the following participant, who was concerned about her illness dominating her daughter’s  
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23 life.  
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27 “I don’t want my daughter having to look after me. She has a full-time job and I just  
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29 don’t want her looking after me. I want her to get on with her life. I don’t want her  
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31 looking after me. She is a dear girl and she loves me very much and I don’t want to  
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33 spoil her life.”  
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37 The participant perceived that the physical burdens would potentially affect her daughter’s  
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39 life by hindering her from fully enjoying her own life. It was understood that the participant  
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41 was concerned about her daughter’s QoL, and she indicated that this would be better if there  
42  
43 were no physical demands placed on her daughter by the need to care for her mother. This  
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45 sense of restricting others’ QoL was perceived as negative. Similarly, another participant  
46  
47 noted how the choice of family holiday destination was affected by ALS:  
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52 “It has to be around me probably. I have to think about myself more there than the rest  
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54 of the family – that gets to me a little bit because I feel they get left out a bit”.  
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3 In addition to these restrictions potentially affecting QoL of significant others, symptoms of  
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5 ALS were perceived to generate psychological burdens on significant others. For instance,  
6  
7 one patient shared how his wife was worried about leaving him alone following multiple falls  
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9 in the house:

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13 “She is afraid of going out unless there is somebody at home to sit with me, so they  
14  
15 keep an eye on me because if she is out I attempt to do things and I shouldn’t really  
16  
17 because I fall on the floor [laughs]”. (Accompanied)  
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19

20 Psychological burdens were also perceived where symptoms violated the previous norms of  
21  
22 participants, and these were perceived to have generated sadness in significant others: “I  
23  
24 know my wife misses conversation with me although we can communicate enough”. The  
25  
26 direct impact of symptoms was therefore acknowledged. At times, the perceived impact of  
27  
28 ALS was not explicitly stated by participants: “My son rings from Australia. I think it must  
29  
30 upset him listening to me talking” (accompanied). Although it was possible that the  
31  
32 participant perceived a negative impact of her ALS symptoms through the violation of the  
33  
34 previous normality, the perceived distress may also be due to being reminded about her ALS  
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36 through the presence of her speech impairment.  
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41 **Future-oriented burdens.** ALS is an illness where prognosis has some uncertainty despite  
42  
43 being progressive. The following quote captures the perceived impact of this on significant  
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45 others: “My partner was in denial, and I could not inflict pain on him by telling him what our  
46  
47 future would be like and how I would deteriorate”. Thinking about their future with ALS  
48  
49 generated distress in her partner because of inevitable deterioration, so it was blocked out.  
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51 Generally, participants described ALS as degenerative, but they also voiced uncertainty of the  
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53 illness. One participant observed that doctors were not able to tell him and his family what  
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3 was going to happen to him, or when, or how “bad” his ALS would become. Imprecision  
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5 about the prognosis of ALS was perceived to provoke worries in his wife:  
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8 “It’s [ALS] getting her down more than it is me. She is always worrying about what’s  
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10 going to happen in the future. Its effects on her are probably, mentally, she would say  
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12 are more than they are on me”. (Accompanied)  
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16 Participants further indicated that ‘uncertainty’ was experienced by their significant others  
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18 not only in terms of the trajectory of ALS, but also in terms of their life without them in the  
19  
20 future. “I’m sure [wife] will [cope] but it’s the unknown. But, if I do die before her how she  
21  
22 will adjust – I think her fear of losing me must be a natural thing.” The foreseeable loss in the  
23  
24 near future was reported to be visibly distressing to significant others, which in turn provoked  
25  
26 its own upset for patients. This psychological burden was verbally expressed by one  
27  
28 participant: “My husband is upset about it. He’s talking about being left on his own”.  
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### 37 **Minimising the burdens**

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40 Participants were keen to minimise the negative impacts of ALS on their significant others in  
41  
42 three areas: physical, psychological, and financial related burdens.  
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46 In terms of physical burden, participants displayed efforts to avoid requests for help:  
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48 “I’m prepared to sit and wait because I know the demands – [wife] can’t do everything”  
49  
50 (accompanied). Some participants were keen to make an arrangement to be looked after by  
51  
52 professional carers so that they would not become a burden to their significant others. For  
53  
54 instance, the participant who expressed her desire not to spoil her daughter’s life had  
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56 communicated her wish to be looked after at a home where she would receive professional  
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3 care when the needs arose. One participant had already put this arrangement in place by the  
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5 time of the interview:  
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8 “I’m quite concerned about my family. But as I’ve taken steps to make sure that I am  
9  
10 looked after – they can go away any time. They don’t have to think, ‘Oh, Mother  
11  
12 can’t get dressed’”.

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15 As suggested here, the aim of these changes was not for the sake of their needs or comfort,  
16  
17 but for the sake of the well-being and QoL of significant others.  
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21 With regard to perceived psychological burdens, participants displayed their attempts  
22  
23 to minimise these by controlling their emotions. One participant clearly described how ‘self-  
24  
25 control’ was necessary to avoid placing a psychological burden on others:  
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28 Interviewee: I just get on with it. They [family] expect me to be strong and I try to  
29  
30 put out all the advantages they would have, and then they change the  
31  
32 subject and to perhaps talk about the... well recently the royal baby  
33  
34 and the names and things like that, and then they will get happy.  
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38 Interviewer: Okay so you kind of control your thoughts and –  
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41 Interviewee: I control their thoughts – that’s the main thing.  
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44 Another participant also shared how she controlled her emotions to avoid unnecessary  
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46 distress to her significant other:  
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49 “I cried for weeks – at first. Before I had confirmation [of ALS], I’d get up in the  
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51 middle of the night and cry alone because I didn’t want to upset my partner  
52  
53 unnecessarily”.

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56 For one participant, this meant he would not hasten death in order to avoid inflicting  
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58 emotional burden on his wife. “This is the way I feel. But, if it was the other way around, I  
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3 wouldn't want her to do that, to end it". For another, it meant she would not make big  
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5 changes to her house so as to keep the house looking 'normal' for her daughter in order to  
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7 avoid a potential psychological burden. Again, this quote illustrates a participant's greater  
8  
9 concern for significant other's well-being than for her own.  
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13 "I have a daughter. If she wants to bring her friends round, I want everything to be as  
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15 normal as possible for her. So, if I can live with a slight change, then that's enough for  
16  
17 me." (Accompanied)  
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20 The most commonly shared effort to minimise future-oriented burdens was made financially.  
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22 Accepting financial consequences of death, participants generally saw putting their financial  
23  
24 affairs in order as important for their own QoL. As articulated by one participant, and  
25  
26 confirmed by a companion, putting her affairs in order and making a will "So, I don't leave a  
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28 mess behind, leave it organised" was good for her well-being.  
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## 36 Discussion

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38 This study found that *concerns for significant others* is an important component of QoL for  
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40 people with ALS. Participants told us how the well-being of their significant others  
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42 influenced their own psychological well-being. This interpersonal impact, however, was  
43  
44 found to be secondary to the existential value of the significant others. Analysis of semi-  
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46 structured interview data showed how significant others added to participant's QoL, as well  
47  
48 as confirming that generally participants had greater concern for their significant others than  
49  
50 they had for themselves. The analysis showed that 'concerns' is separate from 'standards' in  
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52 influencing QoL among this illness group.  
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3 Although general population studies have found a positive impact of having  
4 significant others in the form of a spouse or cohabitant<sup>58,59</sup>, the significance of such  
5 relationships on QoL has not been fully explored in ALS. Previous research does  
6 acknowledge the importance of psychosocial aspects in relation to QoL, but to date, the most  
7 commonly studied aspect of psychosocial significance has been social support within the  
8 concept of coping<sup>60,61,62</sup>. Thus, the current study provides additional knowledge on why  
9 significant others matter to people with ALS. The current study found that significant others  
10 play an important role in individual's QoL because of their influence on people's  
11 psychological well-being, but more notably because of their existential value on people's  
12 QoL.

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27 Participants were concerned about how their illness may affect their significant others,  
28 and how it could reduce their QoL; participants voiced their concerns about restrictions on  
29 significant others from engaging some activities. Assuming that these curtailed activities  
30 were perceived to contribute to the QoL of the significant others, the restrictions were  
31 perceived as negative. Participants were also concerned with the physical demands of their  
32 illness on others, and that this was dominating the life of particular significant others, further  
33 affecting their QoL. The terminal aspect of ALS was found to add to the psychological  
34 burden on significant others, which is in line with findings about psychological burdens on  
35 significant others of people with terminal cancer<sup>63</sup>. In the current study, this terminal aspect  
36 of ALS was found to further generate concerns about financial security for significant others.

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50 In response to the perceived negative impact of ALS on significant others,  
51 participants tried to reduce unnecessary burdens, which involved utilising external support  
52 and controlling their emotions. The terminal aspect of ALS also prompted participants to  
53 secure their financial affairs for their significant others. By exploring participants' attempts to

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3 reduce perceived burdens on others, the findings showed how participants were prioritising  
4 others' perceived needs above their own needs, reflecting greater concern for others. Whilst  
5 acknowledging the significance of sense of self, the current findings show that participants'  
6 concerns for significant others were chiefly rooted in their care for the others. As such, the  
7 current study argues that the foundation of participants' concerns for significant others and  
8 the perceived responsibilities<sup>31</sup> were willingly fulfilled by the participants, even if it was at  
9 their expense. This close yet separate illness impact on QoL in terms of the concepts between  
10 'concerns' for significant others and 'standards' for sense of self has not been explored  
11 previously. The current findings therefore provide rich information to understand how the  
12 significant others are important in their own right for QoL of people with ALS because of  
13 their existential value.

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Previously, illness burdens on significant others have been studied mainly to understand their impact on the significant others<sup>30,64</sup>. The authors acknowledge the importance of understanding how illnesses are experienced by significant others, however, while acknowledging that there is a close relationship between participants' well-being and significant others' well-being, the present study highlights the importance of exploring how people with ALS perceive the burden on significant others in their support of them.

### Limitations of the study

No information was collected to understand the impact of cognitive impairments on QoL. Although all accounts examined in the current study were valid to explore subjective QoL, the study did not explore the impact of any cognitive changes on QoL. Cognitive changes are recognised as common concomitant of ALS in all stages of the illness<sup>4</sup>, and it is plausible that cognitive changes subsequently challenge an individual's perceived QoL. Cognitive changes in social cognition<sup>4,5</sup> may be particularly noted for their potential impact on a patient's



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3 relationship with their significant others, and further their QoL. Given the current finding of  
4 the importance of significant others to our participants, it may be argued that even individuals  
5 with cognitive impairments [still] care about their significant others; they simply fail to  
6 acknowledge or appreciate others' emotions therefore present little sympathy or empathy.  
7  
8 Further study is needed to explore whether individuals with cognitive changes display their  
9 concerns for significant others differently from those with little change to their cognition. In  
10 consideration of these potential impacts of cognitive impairments, the use of cognitive  
11 screening test for MND would have been beneficial.

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22 A second limitation of the current study is the absence of individuals with respiratory  
23 onset ALS. Essentially this was due to the low prevalence rate of this subtype of ALS.

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### Conclusion

The WHOQOL group<sup>36</sup> identifies four aspects that are important in the process of QoL evaluation. In ALS, previous studies have explored how the illness threatens the outlook of people's lives whilst posing challenges to their previous understanding of the self, causing distress. Our findings add to what we know about QoL – as defined by the WHOQOL group – by exploring the importance of significant others as a relevant aspect in QoL of people with ALS. Participants were concerned about their significant others' well-being because of their invaluable existential value, and it was important that perceived burdens to significant others were minimised. In view of this, the current study agrees with a holistic approach in service provision, ensuring the inclusion of relevant significant others. It is further suggested that clinicians explore the specifics of burdens perceived by patients in order to support them to minimise the burdens for their significant others.

## Declaration of Conflict of Interest

The authors declare that there is no conflict of interest.

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## Appendix 1: An interview schedule

### **Introductory question:**

- Could you tell me what you mean by quality of life? What do you understand by it?

### **Key questions:**

- How do you describe your quality of life now?
- What are the things which positively affect your quality of life? How do they affect your quality of life?
- Has there been anything that negatively affected your quality of life? If so, what was it? (If patient does not voluntarily mention how MND may affect their quality of life then ask how their illness has had its impact on their quality of life, if at all.)
- What has been the most difficult change to cope with MND? Why was/is it difficult?
- How do you deal with things that negatively affect your quality of life?

### **Ending questions:**

- Think back to when you did not have the condition, has your definition of quality of life changed after the illness? If so, how?
- Do you have anything else that you want to mention before we end the interview?