King, Rachel (2021) *Living with Motor Neurone Disease: A meta-synthesis of experiences of non-invasive ventilation and an interpretative phenomenological analysis of the experience of distress.* D Clin Psy thesis.

Living with Motor Neurone Disease: A meta-synthesis of experiences of non-invasive ventilation and an interpretative phenomenological analysis of the experience of distress

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Submitted in partial fulfilment of the requirements for the degree of

Doctorate in Clinical Psychology

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July 2021

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Acknowledgements

A special thank you to the participants of this study, who gave their time to take part in the research interviews to share their experiences with me, and who made this study possible.

A huge thank you to Professor Jon Evans for your invaluable guidance and support. Thanks also to Dr Luke Williams, for your advice and perseverance regarding recruitment. I am grateful to both of you for providing expertise and reassurance throughout training, but particularly during that final stretch.

Thank you to the clinicians within the MND Clinic in Ayrshire and Arran for all of your help with recruitment. Your help was greatly appreciated, especially during a particularly demanding year.

Thank you to Paul Cannon for providing expertise and guidance on developing search strategies for the systematic review. Thanks also to Maria Johnson for all of your help with the systematic review, it was greatly appreciated.

Thank you to my family and friends for their encouragement, support, many welcome distractions and reminders of the importance to take a break! Finally, thank you to my partner for his support and for taking on the load of many small things, which made a huge difference.

Chapter One: Systematic Review

Experiences of Non-Invasive Ventilation for Patients with Motor Neurone Disease and Their Carers: A Qualitative Systematic Review

Prepared in accordance with the author requirements for the International Journal of Qualitative Studies on Health and Well-being (see Appendix 1.1, p.85)

Abstract

Purpose

This systematic review aimed to synthesise qualitative literature exploring experiences of being offered non-invasive ventilation (NIV) as part of treatment in Motor Neurone Disease (MND), from the perspectives of patients and carers.

Methods

A systematic search was conducted to identify qualitative studies that explored experiences of MND patients and carers who have been offered and/or use NIV. A quality appraisal tool was used to assess papers prior to synthesis. Meta-ethnography was conducted on ten research studies.

Results

Five superordinate themes were identified; 1) Attitudes and coping styles; 2) Balancing discomfort with benefits; 3) Impact of continuing deterioration; 4) Fear and uncertainty; 5) Challenges of professional support.

Conclusions

The synthesis identified attitudes and coping strategies that patients use to maintain quality of life, autonomy, and control, and how these can change over time. Uncertainty impacts on both patients and carers, who experience a loss of autonomy which complicates decision making around NIV. Fears played a role in rejecting or withdrawing from interventions, particularly a fear of dependence. Loss of autonomy can be experienced during interactions with professionals. Recommendations for clinical practice are discussed, such as improving individualised information and support; increasing autonomy; and improving person centred care.

Keywords: Motor Neurone Disease, Non-Invasive Ventilation, qualitative, meta-synthesis, systematic review.

Introduction

Motor Neurone Disease (MND) covers a group of neurodegenerative diseases which involve the progressive deterioration of upper and lower motor neurones. This results in a loss of voluntary muscle movement, and subsequently a decline in movement, speech, breathing and swallowing (Young et al., 2019). Changes can also include frontotemporal dysfunction, including changes to cognition and behaviour (Strong et al., 2017). Most people with MND die within 3-5 years after symptom onset, often from health complications related to bulbar dysfunction or respiratory failure (Burkhardt et al., 2017). Although some treatments can prolong survival, their main aim is to improve symptom management and quality of life where possible. Respiratory weakness can result in difficulties which impact on quality of life, such as a lack of concentration, poor sleep, fatigue, and shortness of breath (Lyall et al., 2001).

Non-invasive ventilation (NIV) is an intervention to support breathing which can help control respiratory symptoms. . NIV is a type of assisted ventilation, involving a small machine which provides increased air flow to the lung, usually through a mask which is held on by straps. This type of ventilation is seen as non-invasive in relation to interventions such as a tracheostomy, otherwise known as invasive ventilation. This also provides increased air flow, but involves undergoing surgery to allow a tracheostomy tube to be inserted into the windpipe. NIV can allow people to have this treatment at home, but this usually requires support from family members or professional carers (O'Neill et al., 2012).

NIV has been shown to improve quality of life (Walsh & Murphy, 2020), sleep quality (Boentert et al., 2015) and survival in MND (Burkhardt et al., 2017). According to the NICE guidelines on the assessment and management on MND (2016; updated 2019), all patients with respiratory muscle decline should be offered NIV. The use of NIV has increased as evidence of its benefits have increased and factors such as rapid disease progression are no longer considered as barriers for the intervention (O'Neill et al., 2012). Family caregivers who had relatives with MND who were receiving

NIV were found to be less anxious than those whose relatives were not receiving this treatment (Cousins et al., 2013).

Despite these advantages, not everyone who is offered NIV will accept it. Despite its recommendation in treatment guidelines, utilisation rates of NIV are low (Spittel et al., 2020). Transitions such as adjusting to life-sustaining treatments (e.g., home mechanical ventilation or gastrostomy) and using new communication methods can impact on the quality of life of people with motor neurone disease and their caregivers (e.g., Caron & Light, 2015). Relationships between the patient and caregiver can also be negatively affected (Cipolletta & Amicucci, 2015). These transitions can seem constant, with individuals and families facing complex decisions, often planned far in advance while dealing with day-to-day uncertainty.

A mixed methods study examining factors influencing acceptance of NIV and gastronomy highlighted the complexity of this decision-making process (Martin et al., 2014). Psychological factors such as beliefs and perceptions around MND and interventions such as NIV and gastrostomy, and attitudes towards interventions and disease management, were likely to influence decision making. Demographic factors, illness variables and executive dysfunction also had an impact. Having family caregivers who are able to cope with the transition to NIV has been highlighted as an important factor for enabling MND patients to sustain NIV treatment (Cousins et al., 2013). This may be of increased importance due to lack of support from services. MacLaren et al. (2019) suggested that people do not receive adequate care resources to support the provision of ventilation.

Research on the experiences of using home mechanical ventilation across conditions indicates that patients have the contradictory experience of increased freedom and quality of life alongside increased dependency and uncertainty (Ørtenblad et al., 2019). Some qualitative research focusing on the experiences of non-invasive ventilation within MND has identified similar themes as well as focusing on how these experiences can change over time (Ando et al., 2014a) and how this can influence decision making (Ando et al., 2014b).

A recent study reviewed quantitative and qualitative data on the optimal provision of NIV (Baxter et al., 2019). This provides a comprehensive overview of available research and provided recommendations for clinical practice. However, this review took a mixed methods approach to integrate this data. It would be worthwhile to conduct a quality appraisal of additional available qualitative evidence which may have been out with the scope of the previous review and to carry out a meta-synthesis. Studies which have looked at the experiences of MND patients and carers of being offered NIV have produced rich, descriptive data. Synthesising these findings together as part of a meta-synthesis will provide an in-depth insight into these experiences and complement quantitative data reported elsewhere. Improving our understanding of how people with MND perceive being offered and making choices around NIV, and how they experience any benefits and disadvantages of using NIV may allow healthcare professionals to improve how they support patients and carers in making decisions regarding use of NIV and how they support patients in longer-term use of NIV.

While there have been changes in clinical practice and a wider recognition of the impact of NIV on survival and quality of life, it is important that patients’ experiences of care and treatment are presented alongside both evidence for an intervention and the views of healthcare professionals. Increased understanding of perceived obstacles to NIV and benefits of NIV will help to increase insight into positive experiences of NIV other than survival.

Purpose of this review

The review aimed to synthesise the available evidence exploring the experiences of being offered NIV as part of treatment in MND, from the perspectives of both patients and carers. Study quality was appraised and meta-ethnography was used to generate new insights into patients and carer experiences of NIV.

Method

This systematic review followed the guidance in the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA). A protocol for the review was preregistered with PROSPERO International Prospective Register of Systematic Reviews (CRD42021243672).

Search Strategy

The following search was carried out in March 2021. Search terms and database selection were developed in consultation with a subject librarian. Four electronic databases were searched - CINAHL, EMBASE, Medline and PsycINFO. References and citations were searched within selected studies.

Search Terms

Identified databases were searched using key terms for the three key components: ‘motor neurone disease’, ‘non-invasive ventilation’ and ‘qualitative research’. Key words and indexing terms following search terms (MeSH and thesaurus terms) were used as subject headings and title or abstract terms, which varied between databases. A validated qualitative filter was used in searches where possible and adapted for different databases. Within each component the Boolean operator OR was used to group terms and components were combined with AND (see Appendix 1.1 for a detailed search strategy).

Eligibility Criteria

Studies were included if; i) participants were patients with a diagnosis of MND, or carers of patients with MND; ii) the study was a qualitative or mixed methods design; iii) the focus was on the experience of non-invasive ventilation; iv) the paper was published in a peer reviewed journal; v) the study was in English.

Studies were excluded if; i) participants were all health care professionals; ii) the focus of the paper was not about the experience of non-invasive ventilation or focused solely on invasive ventilation;

iii) participants had a diagnosis other than MND; iv) the study was a quantitative design; v) the study was a review or conference abstract.

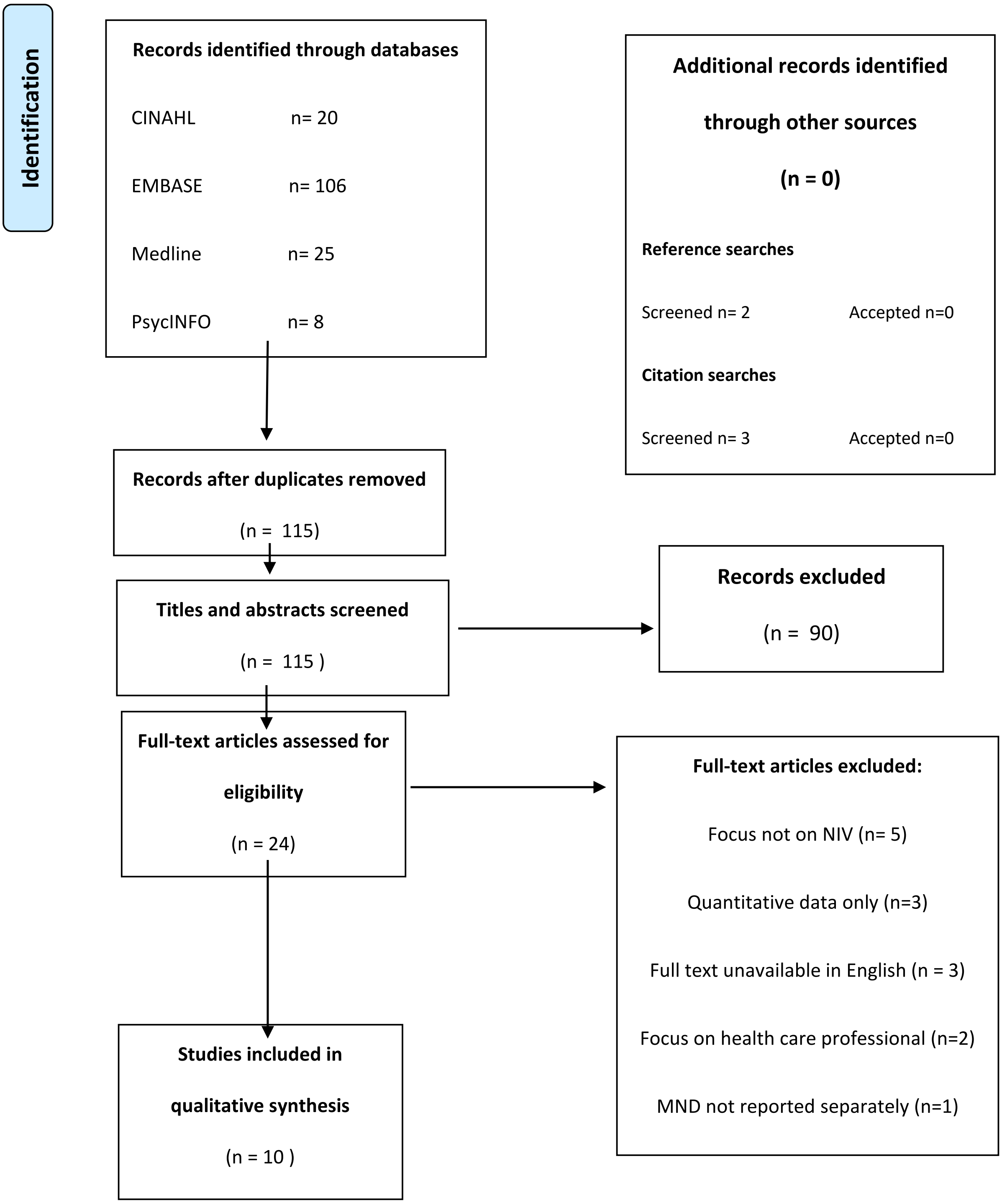
Data Selection

The search yielded 159 results, of which 44 were duplicates (See Figure 1). Electronic searches were documented for transparency (see Appendix 1.2). Titles and abstracts were screened by the primary researcher utilising the inclusion and exclusion criteria, prior to screening the remaining full text papers. A peer reviewer (Trainee Clinical Psychologist) screened 50% of the titles and abstracts and there were no disparities between reviewers. 90 records were excluded at this stage. Hand searches of references, grey literature and citation searches did not identify any additional eligible papers. The full text of 24 papers were reviewed, 10 of which met inclusion criteria for the synthesis.

Two papers (Ando et al., 2014a and Ando et al., 2014b) were part of the same longitudinal study of NIV in MND. The first paper examined experiences of NIV treatment after 12-14 months of use, whereas the second interviewed participants who declined or withdrew from NIV and were therefore not included in the first study. Another three papers (Baxter et al., 2013a; Baxter et al., 2013b; Baxter et al., 2013c) were part of a longitudinal study examining experiences of NIV at different time points and from different perspectives and appears to include the same participants. The question of whether to group these two sets of papers together was considered, but it was decided to include all studies separately as they reported on different aspects of experiences.

Figure 1.

PRISMA Flow Chart



Data Extraction

The JBI QARI Data Extraction Tool for Qualitative Research (Appendix 1.3) was used to standardise the process of recording relevant information from each article.

Quality Appraisal

There is a lack of consensus around how best to evaluate the quality of qualitative research. Majid and Vanstone (2018) reviewed a range of quality appraisal tools for qualitative research and produced a guide to selecting appraisal tools, which was utilised to select the Critical Appraisal Skills Programme (CASP) Qualitative Checklist (2018; Appendix 1.4). This has been identified as appropriate for novice researchers and is a widely used tool with clear guidelines to follow. The researcher and an independent reviewer (Trainee Clinical Psychologist) initially reviewed a sample of two studies, where agreement between reviewers was 80%. Discrepancies were discussed and resolved, resulting in full agreement between reviewers. Both reviewers then reviewed the remaining eight studies, with an agreement of 94%.

The quality rating was not used to exclude studies with a low-quality score as it has been recognised that appraisal may be influenced more by how much detail the authors have been able to describe the methods in and the quality of the report rather than the procedure in qualitative research (Atkins et al., 2008). The potential impact of study quality on findings was considered as part of the synthesis.

Method of Synthesis

Meta-ethnography is a form of interpretative synthesis developed by Noblit and Hare (1988) and is the leading approach to qualitative synthesis within healthcare research (University of Stirling & NHS Quality Improvement Scotland, 2011). It is particularly well suited to integrating data from studies looking at patient experiences and allowing new insights into these, as it allows for the re-interpretation of conceptual data rather than simply providing a description of findings. Meta-ethnography is therefore suitable to synthesise qualitative research exploring the experiences of NIV in MND patients and carers, which will add value to the existing review on this area.

Sattar et al (2021) recently developed a practical guide to conducting meta-ethnography based on the stages set out by Noblit and Hare, in order to improve clarity and guidance for the synthesis process. This paper was used to guide the process of the seven steps of the process of meta-ethnography (see Appendix 1.5). An additional data extraction form was used to record the themes, first order constructs (participant quotes) and second order constructs (primary author interpretations) for each study. Synthesis should ideally involve both reciprocal translation, where themes in one study can incorporate those of another, and refutational translation, which looks to explain and explore differences between studies. Synthesis may also represent a line of argument. Common themes and concepts were then combined across the ten studies to develop third-order constructs (Sattar et al., 2021).

Results

Study Characteristics

Table 1 summarises the ten papers included. Studies explored MND patient and carer experiences of non-invasive ventilation and issues surrounding this intervention. Three studies specifically examined experiences of decision making about NIV (Ando et al., 2014b; Greenaway et al., 2015; Lemoignan & Ells, 2010), with Ando et al. (2014b) focusing specifically on experiences of declining or withdrawing from NIV. Two studies looked at MND patient and carer experiences of NIV (Baxter et al., 2013b; Sundling et al., 2009), while two other studies (Baxter et al., 2013b; Winther et al., 2020) focused specifically on carer experiences. Baxter et al. (2013c) specifically examined carers' and health care professionals' experiences of end-of-life care with MND patients using NIV (although only carer experiences were included in this study). Two studies focused on the perspective of MND patients (Young et al., 1994; Ando et al., 2014a), with Ando et al. (2014a) focusing on how this changed over time.

Participants were all MND patients or carers who had considered NIV as an intervention or had either accepted or rejected NIV. Eight studies were qualitative and two were mixed methodology, with all studies using semi-structured interviews to collect data. Six studies were conducted in the UK, two took place in Canada and the remaining two were conducted in Denmark and Sweden.

Quality Appraisal

The quality of studies was evaluated using the CASP Qualitative Checklist (2018; see Appendix 1.3). See Appendix 1.6 for a summary of responses. All studies provided a clear statement of the aims of the research and why this was relevant. This was not as clearly set out in Young et al. (1994). Qualitative methodology was appropriate in all studies. The research design of all studies was appropriate in addressing their aims. However, four studies did not explicitly justify how they decided which method of qualitative analysis to use (Baxter et al. 2013a; Baxter et al. 2013c; Sundling et al. 2009; Young et al. 1994).

The recruitment strategy was appropriate in all studies. All studies collected data in a way that addressed the research question. However only two studies (Baxter et al. 2013b and Baxter et al. 2013c) explicitly discussed saturation of data. Four studies did not adequately consider the relationship between researcher and participants (Baxter et al. 2013a; Baxter et al. 2013b; Baxter et al. 2013c; Greenaway et al.2015). All other studies discussed collaboration with colleagues, separate research and clinical roles or acknowledged any bias present.

All studies stated that they had received approval from an ethics committee with the exception of Young et al. (1994). This study did discuss consent and ethical standards in research. Three studies did not describe how the research was explained to participants (Greenaway et al. 2015; Lemoignan & Ells 2010; Young et al. 1994) and only half of the studies considered the effects of the study on participants (Ando et al. 2014b; Baxter et al. 2013a; Baxter et al. 2013b; Baxter et al. 2013c; Sundling et al. 2009).

Reports of data analysis were rigorous in the majority of studies, with the exceptions of Winther et al. (2020) and Young et al. (1994). Additionally, another three studies did not adequately discuss their potential bias in the analysis process (Greenaway et al. 2015; Lemoignan & Ells, 2010; Sundling et al. 2009). All studies presented a clear statement on findings, however three studies (Baxter et al. 2013c; Sundling et al. 2009; Young et al. 1994) provided limited critical appraisal. Four studies (Baxter et al. 2013c; Greenaway et al. 2015; Sundling et al. 2009; Young et al. 1994) provided limited or no suggestions for further research. Overall, the quality of studies was generally good, with some areas of exception in Young et al. (1994).

Table 1

Table of Characteristics from Studies in Synthesis

Author (year), country	What the study examined	Design	Methodology	Participants	Main themes
Ando et al. (2014a), UK	How patients' perceptions of NIV treatment evolve over time and how this is reflected in their adherence to NIV	Qualitative	Semi-structured interviews, IPA	N =5, patients with MND using NIV for 12+ months 51-75yo (M=59) 4 male, 1 female	1. Experiences of NIV: physical-psychological relationship 2. Influence on attitude 3. Perceived impact of NIV on diagnosis
Ando et al. (2014b), UK	Why some patients decline or withdraw from NIV	Qualitative	Semi-structured interviews, IPA	N =9, patients with MND who had declined or withdrew from NIV 40-79yo (M=67) 7 male, 2 female	1. Preservation of the self 2. Negative perceptions of NIV 3. Negative experiences with health services 4. Not needing NIV
Baxter et al. (2013a), UK	Whether patient use of NIV impacted on their family carer, and to explore other sources of carer burden	Mixed methods	Quantitative Questionnaires Qualitative Semi-structured interviews, thematic analysis	N= 16, family carers of patients with MND Ages of patients ranged from 'under 60' to '70+', no mean given 12 female cares, 3 male, 1 unspecified	1. Carer perceptions regarding the impact of noninvasive ventilation 2. Carer perceptions regarding sources of burden 3. Role change and patient needs 4. Difficulty having time away 5. Acceptance of professional help 6. Timing of equipment and support services

Author (year), country	What the study examined	Design	Methodology	Participants	Main themes
Baxter et al. (2013b), UK	Experiences of patients with MND and their carers following the recommendation to use NIV	Qualitative	Semi-structured interviews, thematic analysis	N = 20 patients, 17 carers. Patients aged from ‘under 60’ to ‘70+’, M=67) No ages for carers 15 male patients, 5 female. 11 female carers, 3 male, 3 unspecified	1. Potential barriers 2. Perseverance 3. Perceived benefits
Baxter et al. (2013c), UK	Carer and health professional experiences of end-of-life care of motor neurone disease patients using non-invasive ventilation	Qualitative	Semi-structured interviews, thematic analysis	N =9 family carers. Data from HCPs not extracted Patient age: ‘60+’ to ‘70+’, no mean age given. 6 female cares, 3 male	1. Unexpected speed of deterioration 2. Hospitalisation versus dying at home 3. Attempts to resuscitate 4. Decision-making regarding the withdrawal of NIV* 5. Peaceful final moments 6. Turning off the machine 7. Professional uncertainty regarding the use of NIV* 8. Positive impacts of NIV use 9. Concerns regarding NIV use* *These themes were not included in the review as they applied to healthcare professionals only.
Greenaway et al. (2015), UK	Factors associated with decisions made by patients with amyotrophic lateral sclerosis (ALS) to accept or decline non-invasive ventilation (NIV)and/or gastronomy	Qualitative	Semi-structured interviews, thematic analysis	N=21 total, N= 5 who decided about NIV 3 participants were interviewed with carers present. NIV participants: 4 male, 1 female 50-71yo (M=64)	Patient-Centric Factors 1. Perceptions of choice 2. Acceptance and need 3. Aspects of fear* External factors 4. HCPs: Doctors, nurses, therapists, care workers 5. Family* 6. Information 7. The Concept of Time *These themes or subthemes were not included in the review.

Author (year), country	What the study examined	Design	Methodology	Participants	Main themes
Lemogian & Ells (2010), Canada	The experiences of decision-making about assisted ventilation for ALS patients	Qualitative	Semi-structured interviews, thematic analysis	N =9 total N using/making decisions about NIV = 8 NIV patients: 5 male, 3 female 36-72yo (M=54)	1. The Meaning of the Intervention 2. The Importance of Context 3. The Importance of Values 4. The Effect of Fears 5. The Need for Information 6. Adaptation To or Acceptance of the Intervention Quotes/interpretations from patients solely using long term mechanical ventilation were excluded
Sundling et al. (2009), Sweden	Patients with ALS and their caregivers' experiences of non-invasive positive-pressure ventilation	Qualitative	Semi-structured interview, content analysis	Patients N=7, 5 male and 2 female 45-73yo (no mean given) Caregivers N=8, 6 female and 2 male 40-74yo (no mean given)	Patient Experiences 1. Getting to know the ventilator 2. Embracing the ventilator 3. Being on a ventilator on a 20-24-h basis Caregiver's Experiences 1. Getting to know the ventilator 2. Embracing the ventilator 3. Being on a ventilator on a 20-24-h basis
Winther et al. (2020), Denmark	Everyday life experiences of relatives of people with amyotrophic lateral sclerosis (ALS) living at home with mechanical ventilation and formal caregivers	Qualitative	Semi-structured interviews, structural analysis	N=11 total, N=9 carers of patients using non-invasive mechanical ventilation only/at some point in illness Carers – 5 female, 4 male Ages not specified	1. We are in this together until the end 2. Vulnerable relatives fighting to keep a track of everything 3. Formal caregivers – a distressing relief 4. A prison without personal space Quotes and interpretations were only included from patients who were receiving NIV or had done in the past

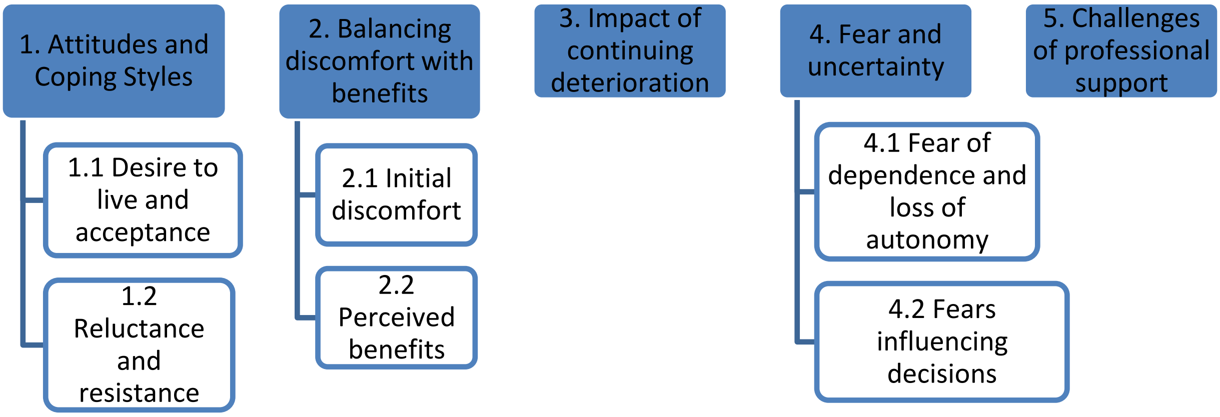
Author (year), country	What the study examined	Design	Methodology	Participants	Main themes
Young et al. (1994), Canada	Issues of the use of mechanical ventilation from the patients' perspectives	Mixed methods	<u>Quantitative</u> Factors rating scale to identify areas of importance, questionnaires <u>Qualitative</u> semi-structured interviews based on factors, thematic analysis	N=13. 11 older than 55yo, 2 younger than this (no mean provided) 11 male, 2 female 4 of 13 participants were using mechanical ventilation, with 3 of these being on NIV The other participants were being asked to considered mechanical ventilation	1. Quality of life 2. Severity of disability 3. Availability of ventilation by means of a nasal mask 4. Possible admission to a long-term care facility 5. Ability to control discontinuation of ventilation 6. Desire to live 7. Concerns for the emotional health/burden of care of family members 8. Feelings about the ventilator Any quotes specific to invasive mechanical ventilation (unless being compared to NIV) were excluded

Meta-Synthesis

The meta-ethnography process elicited 5 super-ordinate themes through reciprocal translation (see Appendix 1.7); 1) Attitudes and coping styles; 2) Balancing discomfort with benefits; 3) Impact of continuing deterioration; 4) Fear and uncertainty; and 5) Challenges of professional support. The five superordinate themes and subordinate themes are detailed in Figure 2. All themes which emerged from lower quality studies were consistent with themes which emerged from studies of a higher quality.

Figure 2

Superordinate and subordinate themes



1. Attitudes and coping styles

The majority of studies reported on patient and/or caregiver attitudes towards MND and NIV, and how these influenced coping styles.

1.1 Desire to live and acceptance. The importance of living was highlighted by many patients, with NIV seen as essential to survival. Some patients viewed this essentiality as reflective of a lack of choice (e.g., Greenaway et al., 2015). Acceptance of MND appeared to be linked with acceptance of interventions as patients wished to live for as long as they could. It was highlighted in Lemogian & Ells (2010) that acceptance is a gradual process of repeated adaptation, involving familiarisation with equipment. The importance of perseverance was highlighted in a couple of papers; if patients experience that it allows them to live their life as they would want to, this can increase acceptance (e.g., Ando et al. 2014). Coping strategies such as problem solving and living in the moment appeared to be linked to psychological benefits of NIV and facilitate further adaptations such as acceptance of support. Caregivers reported acceptance of the disease, allowing them to come alongside patients as a collective "*we*".

"I would like to live and living (sic) as long as I can". Patient (Ando et al., 2014a)

"We're dealing with things as we have to, and it's probably not a bad way of going about it. And just thinking I'll worry about it when it gets here. I can't worry about it before." Patient (Greenaway et al., 2015).

1.2 Reluctance and resistance. Resistance to accepting MND seemed to result in patients being less likely to accept NIV, for the same reason that people did accept it: a desire to maintain normality as much as possible, with acceptance of NIV being seen as giving into MND. These patients viewed NIV as prolonging an inevitable death. Ando et al. (2014a) found that there can be a shift in this attitude over time for some patients, particularly if they experience benefits of intervention. This view appears to reflect a resistance to perceived changes in identity and independence, and a desire to maintain control. Resistant patients equated preservation of life to a loss of dignity, dependence on others and an unacceptable quality of life. While they expressed a desire to live, living with NIV was seen as *"not really living"*. Some resistant patients perceived that they did not require assistance, despite noticing changes to their breathing.

"I've got a strong desire to live...but not with a ventilator." Patient (Young et al., 1994)

2. Balancing discomfort with benefits.

Most studies reported on the initial discomfort of using NIV, and/or the benefits that patients and caregivers experienced from its use over time.

2.1 Initial Discomfort. It was common for patients to describe a negative first impression of using NIV. There are various aspects of NIV which can cause discomfort; the mask and strap, mask sores and lesions were identified as ongoing difficulties.

A common source of distress was air pressure, with this sensation feeling too powerful or making patients feel like they could not breathe. Mouth dryness was also common. Initially, patients and carers may feel as if life is dominated by the ventilator until this becomes integrated into their routine. Carers and patients experienced an initial issue with noise and/or interrupted sleep. This alongside the responsibility of motivating patients to use the ventilator, an initial lack of familiarity with the ventilator and frustration around its functioning can result in negative emotions. It appears that many disadvantages become easier with continued use, but some discomfort persists.

"It did seem to take over, that became extremely important. But as the time's gone on, as I say, we've got a lot more relaxed with it.'' – Carer (Baxter at al., 2013a)

2.2 Perceived benefits. Improved sleep was the most commonly reported physical benefit for both patients and carers. Patients also reported improved energy levels, reduced shortness of breath and that it maintained their ability to communicate. These factors impacted on psychological wellbeing as patients had more energy to engage in valued activities e.g., visiting family, carrying out daily activities and living at home. NIV was generally perceived as improving quality of life as a result. NIV was described as eliciting feelings of relief and relaxation. Patients identified that a benefit of NIV was that it was a "*no risk*" decision as it was non-invasive and easy to discontinue, making it more acceptable than tracheostomy.

Carers experienced similar feelings of relief reported by patients. Carers described NIV as being easy to use and maintain. Studies found that carers benefitted from seeing the positive impact of NIV on patients and encouraged its use. They experienced less anxiety around the patients breathing and had more short periods of time to complete activities for themselves. In one study, carers reported a peaceful death for patients on NIV, with no choking or struggling for breath (Baxter et al., 2013c). Some carers were unsure about whether the minimal benefits their relatives received from NIV was worth the effort, but as a whole NIV was commonly experienced as beneficial for everyone.

''The whole family is at peace. Everything's changed. He's safe and I can relax. It's…almost…perfect.''

Carer (Sundling et al., 2009)

3. Impact of continuing deterioration

The course of MND was often experienced by patients as unpredictable, making decision making around interventions for both the present and the future difficult. This could be perceived as having little or no choice when accepting an intervention such as NIV, as it became a necessity. Conversely the uncertainty of decline and its impact in MND made others completely unable to accept or reject and intervention. Deterioration appeared to result in increased awareness of disease progression for some patients, which impacted on decision making.

"…with ALS there's no ruler.. .to measure where you're at. . .. It's not being able to know what's normal or what's next." – Patient, Lemogian and Ells (2010)

Increasing deterioration was associated with changes in roles for both patients and carers, with more responsibility placed on caregivers in terms of practical tasks and physical demands. NIV itself did not appear to be a significant contributor to carer burden. Carers experienced changes as being so gradual that they were difficult to identify. Winther et al. (2020) highlighted behavioural changes can be even more difficult for carers than practical challenges.

"I didn't realise how quickly he would go down, I really didn't. But I'm thankful that he did." – Carer, Baxter et al. (2013c)

4. Fear and Uncertainty

Almost all studies touched on fears that patients experience about using NIV, and how the uncertainty of MND influences decision making.

4.1 Fear of dependence and loss of autonomy. Being in control of their condition and decision making was important to patients. Some patients felt that they had little choice around accepting an intervention, either due to healthcare staff or because of the disease itself.

A common experience for patients across studies was the fear of becoming dependent on NIV, with some patients viewing acceptance of NIV as giving in, or being concerned about NIV causing more rapid deterioration. Ando et al. (2014b) found that this view changed for some people after using NIV for a period of time. NIV could be seen as a loss of autonomy and therefore a challenge to one's identity and resulting in a lesser quality of life, both which could be viewed as unacceptable. This is contradictory to patients who reported one benefit of NIV as choice and feeling in control e.g., a choice to discontinue.

Carers experienced loss of autonomy, with less control and choice over their own lives as the disease of the patient progressed. Carers also emphasised the importance of feeling involved and in control of decision making around interventions.

"The main thing is being totally dependent on a machine and not being able to control my own destiny." Patient, Young et al. (1994)

4.2 Fears influencing decisions. Feelings of claustrophobia were a common potential issue of NIV use identified across studies, and patients identified worrying that NIV use was unbearable or unnatural. Other fears were broader, such as fear of deterioration, death and the future, and the uncertainty surrounding these. Some patients feared having no other choice than to use NIV.

"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." – Patient, Ando et al. (2014b)

5. Challenges of professional support

Some patients described difficult experiences with healthcare professionals (HCPs) and felt the service they received was poor. For some patients this was linked with disengagement and refusal of interventions. Some patients experienced HCPs as persistent about interventions, which could be seen as a threat to autonomy. Overwhelming numbers of appointments and feeling like there was no other choice than to trust the expert also added to this sense of threat. Others felt that HCPs were supportive and helped with the decision-making process by providing reassurance. Winther et al. (2020) found that carers could feel neglected and undervalued by HCPs.

"…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." Patient, Ando et al. (2014b)

Carers reported experiencing a burden of responsibility for both providing care and for coordinating between multiple agencies. Support from professional carers was difficult for carers to accept and presented additional challenges. Carers felt a responsibility to train and support professional carers, and found it difficult to trust their loved one's care to others. Common experiences were worry, guilt, lack of privacy and frustrations over communication issues. These additional stressors appeared to become increasingly challenging as patients deteriorated.

"….it is a great invasion – that suddenly there are people in your home all the time" – Carer, Winther et al. (2020)

Discussion

This systematic review examined available evidence exploring the experiences of being offered non-invasive ventilation as part of treatment in Motor Neurone Disease (MND), from the perspectives of both patients and carers. The synthesis identified five superordinate themes; 1) Attitudes and coping styles; 2) Balancing discomfort with benefits; 3) Impact of continuing deterioration; 4) Fear and uncertainty; and 5) Challenges of professional support.

Acceptance appeared to be linked to positive coping strategies. Findings suggest that the acceptance of NIV was related to the acceptance of having MND, but recognised that this is a complex and dynamic process. There can be a change from resistance to acceptance over time. This may reflect the ‘response shift’, a concept first integrated into health related quality of life research by Sprangers and Schwartz (1999). The response shift has been used to explain why quality of life in progressive conditions such as MND can remain relatively high and stable. The response shift is an important part of how people adapt to a chronic illness, involving the mechanisms of recalibration, reprioritisation and reconceptualisation to bring their expectations in line with reality (Sprangers & Schwartz, 1999). In MND, people face continuing deterioration and therefore ongoing adaptation to what they are able to do independently, with support or not at all. Simmons et al. (2015) highlighted that this deterioration can result in a change from physical functioning having the biggest impact on quality of life, to other factors unrelated to physical functioning which are important to people (e.g. family, friends, interests and purpose). Oh et al. (2021) reported that avoidant strategies were commonly used in earlier disease stages of MND, while acceptance was associated with later stages. This review suggested that the utilisation of coping strategies is a constant process through the disease course as people adapt to both short- and long-term stressors. Lazurus and Folkman’s (1984) stress and coping theory differentiates between emotion focused and problem focused coping. They suggest that coping is situation dependent, with emotion-focused strategies used when situations feel more uncontrollable (e.g., with increased deterioration) whereas problem-focused strategies are used in more controllable situations (e.g., when people learn more about the impact of such deterioration).

This shift may influence whether patients are able to focus on the discomfort caused by an intervention such as NIV or on the benefits over time. It was common for patients and caregivers to focus initially on how uncomfortable NIV can be, with benefits appreciated over time as patients saw how it could help them to engage in valued activities. A scoping review of supportive care needs of MND patients and carers (Oh & Kim, 2017) identified that practical needs (including with assisting devices such as NIV) were most frequently reported. This was linked to informational needs, such as education around disease progression, treatment, and home-based caring skills. Education was found to reduce carer burden and information on what to expect with NIV may be beneficial for patients and carers and help them to persevere with treatment. Greenaway et al. (2015) highlighted the importance of information being individualised and coming from a variety of sources, while Baxter et al. (2013b) found a preference for in person or telephone support over written information. Patients in Lemoignan & Ells (2010) wanted ongoing information as the disease progressed.

Disease progression brings uncertainty for patients and carers, which can make decision making difficult. Role changes also occur, with carers experiencing increased responsibility. The experience of uncertainty has been documented in MND, beginning when people are seeking a diagnosis (Remm et al., 2019). Uncertainty has also been identified as a difficulty for carers, who need to constantly adjust to changing demands including assistive interventions and anticipating equipment needs (Anderson et al., 2019).

Fears that NIV use would be claustrophobic or unnatural were identified. For some patients, it appeared that assistive technologies further forced them to confront their own mortality. Fear of losing control and independence was the most prominent concern. Hobson et al. (2017) identified resistance to specialist assistive technology which led to avoidance of thinking about or accessing this support even if this was seen as useful. Requiring specific interventions can be perceived as reaching unwanted milestones. It is important for people to maintain control and autonomy where possible while living with a disease which removes this. It may be that people maintain autonomy through acceptance and preparation for the future, or by avoidance and focusing on the present. Soundy & Condon (2015) found that autonomy influenced wellbeing across different stages of the disease. One element of this was difficulty in accepting interventions and devices as this could be seen as giving into MND, which can influence how people view themselves.

The importance of autonomy was linked to how patients and carers experienced professional support, as it was important for people to feel involved in decision making. Some patients reported the experience of services as poor, which could influence whether patients accepted interventions. Remm et al. (2019) found that poor experiences with HCPs could increase chances of disengagement. Patients also reported HCPs were persistent about interventions, limiting their own autonomy. Carers felt that responsibility was placed on them without recognition from HCPs. Additionally, support from carers came with a loss of choice around privacy. Higher levels of perceived control in caregivers have been found to predict lower caregiver distress (de Wit et al., 2020).

Limitations

No carers in the included studies reported challenges around coping with behavioural and emotional changes in MND patients, with the exception of one participant in Winther et al. (2020). Insight into the difficulties that carers may face when making decisions about interventions such as NIV and in supporting the implementation of interventions at home when their relative has frontotemporal dysfunction/dementia would have been valuable. These difficulties are being increasingly recognised in MND, with behavioural changes reported as a source of caregiver burden (e.g., Andrews et al., 2016).

This review was limited to studies published in English; therefore, relevant papers may have been missed as two papers were excluded on this basis. The papers included span a period of 26 years, during which NIV will have changed considerably. Although studies were primarily conducted in the UK, studies from other countries were included where healthcare services may be experienced differently. However, themes were evident across studies and implications for services are important to highlight regardless of whether these are in place or not.

Quality appraisal was conducted to highlight any limitations rather than to exclude studies from the synthesis. Lower quality studies may have influenced findings. However, themes in these studies were comparable to themes found in studies of a higher quality.

Only information related to non-invasive ventilation was extracted from studies. Therefore, some information may have been missed. It would be useful for further studies or reviews to examine the difficulties that come between choosing between NIV and invasive ventilation (e.g., tracheostomy) or in progressing from invasive ventilation from NIV, as this experience is likely to come with additional challenges.

Baxter et al. (2013a;2013b;2013c) likely interviewed the same participants across studies, therefore themes are likely to have been repeated. However, only one subordinate theme draws from all three of these papers, suggesting that there was variation between experiences.

Implications

Aspects of fear and uncertainty appeared to be exacerbated by lack of information to support patients and carers in their decision making. Patients can feel overwhelmed by the provision of information and support immediately after diagnosis (Soundy & Condon, 2015) highlighting the need for information to be provided as the disease progresses. Baxter et al (2019) highlighted the importance of early NIV intervention, and the challenges of implementing these findings. Patients are often resistant to interventions initially, but attitudes may change over time. Offering support and training may help carers to feel like their needs and burden of responsibility are valued by healthcare professionals. Bergin & Mockford (2016) suggested that carer coping and quality of life can be enhanced when resources are shared between HCPs, carers, and patients.

Results suggest that the experience of healthcare services can influence decision making. Additional research on what could improve patient experiences of healthcare could help to further inform this. Access to accurate information can help people to feel informed in their decision making, which in itself has been found to help build trust and engagement with HCPs in oncology settings (Campbell et al., 2010). Conversely, when patients feel they are not valued and involved in decision making this results in disengagement and potentially rejection of life-sustaining interventions. Exploring the holistic needs of individuals and carers, and framing benefits of NIV in terms of what has been identified as important to people (e.g., quality of life and valued activities) may increase the likelihood of patients accepting a NIV trial. See Table 2 for a summary of practical implications for health care professionals.

Future Directions

Further research on awareness of different coping styles and attitudes may help to identify suitable psychological interventions for patients and carers. A mindfulness-based stress reduction (MBSR) meditation program tailored to MND patients resulted in significant improvements in anxiety, depression, and quality of life both post-intervention and at follow up (Pagnini et al., 2017), with similar results found in an online mindfulness intervention for MND patients and caregivers (Pagnini et al., 2021), where caregivers reported lower levels of carer burden.

A meta-analysis of interventions for family caregivers of cancer patients emphasised the importance of patients and caregivers receiving support simultaneously, since they will react to the diagnosis of cancer as a unit (Northouse et al., 2010). The impact one has on the other was highlighted, and that carer wellbeing provides patients with optimal care. Parallels can be drawn with this experience and MND where patients and families experience the illness and its challenges together.

Conclusions

Attitudes towards MND and NIV and related coping styles appear to be driven by the same goals; to maintain autonomy and control; a desire to live; maintaining an acceptable quality of life; not ‘giving in’; and maintaining self-identity. There may be a shift from resistance to acceptance in the long term, and while individuals deal with ongoing stressors. There may also be a shift from focusing on discomfort related to NIV, to focusing on the benefits. Uncertainty impacts on both patients and carers, who experience a loss of autonomy which complicates decision making around NIV. Fears played a role in rejecting or withdrawing from interventions. Loss of autonomy can also be experienced during interactions with professionals, resulting in disengagement.

Recommendations for clinical practice focused on improving person centred care for both patients and carers. This review has focused on the experiences of MND patients and caregivers who have been offered or have used NIV, with the hope that by improving HCPs understanding of this experience we can provide better support to MND patients and families.

Table 2.

Summary of practical implications for Health Care Professionals

Area	Specific practical implications/recommendations
Information provision	• Information should be individualised and tailored to the individual. • Information should be provided at point of diagnosis – but this can be overwhelming and patients and carers may need to have this explained again. • Patients and carers may need time to process information and to formulate questions. Having access to reliable printed or online information may help. • Information should be provided on an ongoing basis, with opportunities for patients and carers to ask questions. • Information should be provided on what to expect when beginning NIV in terms of discomfort, and on the potential benefits of using NIV.
Education and support	• Providing information or training sessions to carers around how to operate NIV machinery may help to reduce initial anxiety and carer burden. • Ongoing support should be offered to both patients and carers, in general and when using an intervention. If services are unable to provide this, signpost to relevant helplines. • Providing support to patients and carers in parallel may help to improve patient outcomes. • Assist in reducing discomfort as much as possible e.g., support with mask fitting.
Autonomy	• Patients should be supported to make their own informed decisions. • Patients should be offered choice of interventions on an ongoing basis, as their attitudes may change over time. • The provision of accurate information can help patients to feel more involved in decision making about their care.
Relationship with HCPs	• If patients feel that they have autonomy in decision making and are well informed, this may help to build engagement. • HCP understanding that attitudes and perceptions of interventions may change over time may help them to provide more person-centred care. • Acknowledging the demands on carers may also help to improve relationships with patients and carers. • Exploring the holistic needs of patients and carers and explaining the benefits of NIV in terms of what is important to them may increase acceptance of NIV. E.g., impact on quality of life rather than survival.

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Chapter Two: Major Research Project

Experiences of Distress in Motor Neurone Disease and Views on an Adapted Distress Thermometer and Concerns Checklist: An Interpretative Phenomenological Analysis

Prepared in accordance with the author requirements for the International Journal of Qualitative Studies on Health and Well-being (see Appendix 1.1, p.85)

Plain English Summary

Background

The Distress Thermometer (DT) and accompanying Concerns Checklist (CC) are tools which have been shown to be useful for screening distress and supporting health conversations across a range of health conditions (Snowden et al., 2011). Motor Neuron Disease (MND) covers a group of uncommon diseases that affect the brain and nervous system, and which involve muscle weakness which continues to become worse over time. People can also experience changes in their cognition, behaviour and communication abilities. It is important that distress is monitored in MND, as needs change quickly and patients have to adapt to a decline in their abilities, an increase in their dependence on others and changes across all domains of their lives (Sakellariou et al., 2013). Whilst the experience of psychological distress in people with MND is considered to be understandable it remains poorly understood and previous research has not examined how people with MND view DT and CC.

Aim

This study aimed to explore the lived experience of, and the meanings associated with, distress for people with MND. It is hoped that the results of this study may help to improve the understanding of distress in MND and help us to understand how people with MND experience the use of the DT and CC in screening for distress.

Method

A small number of people with MND (n=6) took part in semi-structured interviews. These interviews were then transcribed, and the data was analysed using a technique called Interpretative Phenomenological Analysis.

Results

There were six main themes identified from the analysis; 1) diagnosis related distress; 2) support influencing distress; 3) the distress of uncertainty; 4) loss of independence and roles; 5) coping with MND and distress; and 6) views of the MND tool.

Conclusions

Participants discussed how difficult it is to deal with ongoing uncertainty and multiple losses experienced in MND. They spoke about the impact that personal and professional support can have on distress. Participants reported coping by focusing on positives of the past and present. Findings highlight the need for person centred care, which can be improved by the use of tools to screen for distress and identify concerns in MND. Recommendations which may increase person centred care and the identification of distress are discussed, such as providing information tailored to individuals; increasing awareness of MND in non-specialist health services; and training healthcare professionals to use the DT and CC.

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Abstract

Purpose

To explore the lived experience of, and the meanings associated with, distress in the context of Motor Neurone Disease (MND).

Methods

Phenomenological approach to understanding the experience of distress in MND and whether the Distress Thermometer (DT) captures this experience. Semi-structured interviews were conducted with people with MND (n=6) who were recruited from an MND clinic.

Results

Six superordinate themes emerged from the Interpretative Phenomenological Analysis: 1) diagnosis related distress; 2) support influencing distress; 3) the distress of uncertainty; 4) loss of independence and roles; 5) coping with MND and distress; and 6) views of the MND tool.

Conclusions

Findings highlighted the pervasive nature of uncertainty, multiple losses in MND and the impact that personal and professional support can have on distress. Participants reported coping by focusing on positives of the past and present, and on maintaining normality. Findings highlight the need for person centred care, which can be improved by the use of tools to screen for distress and identify concerns in MND. Recommendations which may increase person centred care and the identification of distress are discussed, such as providing individualised information; increasing awareness of MND in non-specialist services; and training healthcare professionals to use the MND DT.

Keywords: Motor Neurone Disease, distress, distress thermometer, Interpretative Phenomenological Analysis

Introduction

Psychological distress is commonly used as an indicator and outcome of mental health, but the concept remains unclear as it is applied to various symptoms, behaviours, and difficulties (Drapeau et al., 2012). Distress has been defined in a healthcare context as ‘a multi-factorial construct involving a complex interplay between biological, psychological, interpersonal, social, and existential factors’ with an emphasis on taking all of these aspects into account (Brennan et al., 2012, p. 1346). It has been proposed that the term can encompass general worry, fears, sadness and spiritual or existential crises as well as mental health problems (Holland & Bultz, 2007). Measurement of distress has been recognised as important across different clinical populations and health care settings due to the negative impact it can have on outcomes such as treatment seeking, quality of life, treatment adherence and survival rates (Ransom et al., 2006; Donovan et al., 2014; Faller et al., 1999; Holland & Alici, 2010).

While often overlooked within physical health care settings, distress has long been recognised as the sixth vital sign within oncology care, with 25-60% of cancer patients reporting experiencing distress when asked (Zabora et al., 2001). The National Comprehensive Cancer Network (NCCN) developed the Distress Thermometer (DT; initially developed by Roth et al., 1998) and accompanying Concerns Checklist (CC), and advocate for the routine screening of distress. The DT is a visual screening tool where individuals are asked to rate their level of distress over the past week using a 0 (no distress) to 10 (extreme distress)-point Likert scale. Alongside this, the CC allows patients to indicate the source(s) of their distress under categories such as physical, practical and relationship problems.

These tools have been shown to be useful for screening distress and supporting health conversations across a range of health conditions in clinical practice (Snowden et al., 2011). Brennan et al. (2012) highlighted the importance of using tools as part of a structured, collaborative discussion in order to effectively identify and reduce distress in patients. Healthcare conversations contribute to quality of patient care (Hemsley et al., 2015). It is particularly important to ensure that communication is effective when people experience communication difficulties as these individuals often do not receive support that they require (Wylie et al., 2013). One study found that both clients and professionals found preparation and structure helpful in reaching a shared understanding, with which tools such as the DT and CC may assist (Stans et al., 2018).

Various studies have established the validity of the DT as a tool to detect distress in cancer patients by comparing the DT to other screening measures (e.g., Ma et al., 2014; Gessler et al., 2007). A scoping review (Sousa et al., 2021) identified misuse of the DT in non-cancer populations, with 66% of studies using the DT without a concerns checklist, which contradicts recommendations for its use (Cutillo et al., 2017). As a result, the clinical utility of the DT in non-cancer populations remains unclear. Sousa et al. suggested that it was reasonable for concerns checklists to be adapted to specific conditions, and that the DT was both an appropriate and acceptable screening tool for distress across conditions. The DT is currently recommended as a screening tool for epilepsy (Scottish Intercollegiate Guidelines Network, 2018) and chronic respiratory disorders (e.g., COPD; Makuch et al., 2020).

MND covers a group of neurodegenerative diseases which primarily involve the progressive deterioration of motor neurones, which control voluntary muscle movement e.g., movement, speech, breathing and swallowing (Young et al., 2019). These impairments often lead to increased dependency, changes to lifestyle and relationships (Trail et al., 2009). Neuropsychological deficits affect over 50% of people with MND, and these can vary considerably. Revised criteria for frontotemporal dysfunction MND have been expanded to incorporate the concept of a frontotemporal spectrum disorder to accurately reflect changes in executive function, language, memory, and behaviour which can occur in MND (Strong et al., 2017). Little is known about distress related to these changes as many studies focusing on wellbeing have excluded participants with cognitive or behavioural changes; however, behavioural changes have been linked to hopelessness in patients and increased caregiver burden (Caga et al., 2019).

People with MND experience a wide range of psychological reactions, such as anxiety, hopelessness, and distress (Averill et al., 2007). Depression and psychological distress have been found to be unrelated to disease progression or time since diagnosis (Rabkin et al., 2005). Psychological distress can reduce survival rates and quality of life, and increase interest in hastened death (McDonald et al., 1994; Johnston et al., 1999; Ganzini et al., 2002). The experience of psychological distress in people with MND remains poorly understood. Standardised measures examining distress often fail to address patients’ holistic needs, instead focusing on depression and anxiety.

It is important that distress is regularly monitored in MND, as patients and their families face challenges in coping with uncertainty on a daily basis. Needs change quickly with existing symptoms progressing as new ones develop, as patients continuously have to adapt to a decline in their abilities, an increase in their dependence on others and changes across all domains of their lives (Sakellariou et al., 2013). Bulbar symptoms lead to impairment of speech and affect up to 30% of people with ALS (Haverkamp et al., 1995). Communication difficulties change over time and have a significant impact on quality of life; it has been identified by MND patients as a key factor in maintaining or losing their independence (Gruis et al., 2011; Green et al., 2013).

In line with the National Institute for Health and Care Excellence (NICE, 2016) guidelines, which state that the psychological and emotional impact of MND should be discussed during assessments, The Motor Neurone Disease Clinic (MNDC) in NHS Ayrshire and Arran (NHS A&A) developed and introduced the DT with an adapted Concerns Checklist (CC; Appendix 2.1) in 2017 to be administered routinely at appointments. This tool allows patients to address physical, cognitive, emotional, practical, and social needs, and ensures that clinicians enquire about distress.

However, there are additional challenges to identifying psychological and emotional needs in the MND population due to communication difficulties and cognitive impairment. MND services that have audited the use of adapted versions of the DT and CC have found that these tools have allowed patients to identify unmet needs and prompted further referrals (Wright et al., 2011) and that these were acceptable to both patients and clinical staff (Lennox et al., 2018). However, individuals may experience screening differently. A systematic review found that when screening identified distress in cancer patients but provided no follow up care, there was no benefit of screening (Mitchell, 2013). This was in contrast with studies which demonstrated improved patient outcomes when screening was linked with mandatory referral or intervention. Stewart-Knight et al. (2012) emphasised the importance of conducting qualitative interviews in order to understand experiences of distress, what factors contribute to distress and how this is reflected in the DT. This is likely to be a particularly useful approach when the experience of distress within a particular disease, such as Motor Neurone Disease (MND), is not well understood.

Aims

The aim of the research was to better understand the lived experience of, and the meanings associated with, distress in the context of MND. It also aimed to understand patient views on the DT and CC for MND.

Research Questions

i) What are MND patient's experiences of distress, if any?
ii) What is the meaning of distress for individuals with MND?
iii) What are MND patient's views on using the Distress Thermometer and Concerns Checklist in relation to their own experience of attending an MND clinic?

Method

Ethical Approval

This study was approved by the Research and Development Department in NHS Ayrshire and Arran (reference number CM/KLB/CI 2020AA070; see Appendix 2.2) and NHS West of Scotland Research Ethics Committee (reference number 20/WS/0157; see Appendix 2.3). All participants gave verbal consent after receiving the consent form (Appendix 2.4) prior to interview, which was recorded before commencing interviews. Relevant data protection regulations were followed to ensure study data was stored securely.

Design

A qualitative design utilised Interpretative Phenomenological Analysis (IPA) methodology to explore and analyse the lived experiences of people with MND. Participants took part in a semi-structured interview.

Recruitment

The MND team were provided with information about the study (Appendix 2.5), identified eligible participants and informed them about the study. Those interested consented to their contact details being passed on via an opt-in form to the researcher (Appendix 2.6), who then contacted them to discuss the details of the study, what participation would involve, and to arrange a suitable interview time. Participants were sent the Participant Information Leaflet (Appendix 2.7), instructions on how to access Attend Anywhere, a copy of the consent form for their information and the distress thermometer and concerns checklist. Participants were asked to look over the DT and CC prior to interview. Participants were not previously known to the researcher.

Participants and Sample Size

Six participants were recruited in NHS Ayrshire and Arran at the MND clinic within the Douglas Grant Rehabilitation Centre. One other person agreed to be interviewed and sent information, but phoned to cancel their interview for reasons unknown. Inclusion criteria were Diagnosis of Motor Neurone Disease; registered and seen within the Motor Neurone Disease Clinic; cognitive capacity to communicate informed consent; aged 18 years or older, ability to verbally communicate. Exclusion criteria: inability to communicate consent or lack of cognitive capacity to give consent as identified by the clinical team; communication difficulties which would be considered too difficult to understand via audio recording.

The method of IPA is typically based on, and is best suited to, small sample sizes involving individual interviews of participants. In IPA research purposive sampling is normally used in order to have a homogenous sample. This approach was used as much as possible given the small population of people with MND. The study sought to recruit 6-8 participants as this is the recommended sample size for doctoral level research projects (Turpin et al., 1997). Due to constraints on recruitment such as a low number of clients meeting inclusion criteria, data saturation was not considered.

Data Collection

Interviews were carried out between April 2021 to June 2021. One interview was conducted with each participant by the primary researcher (R.K.) Participants were given the option of having multiple, shorter interviews if required. No other individuals were present during interviews and no repeat interviews were carried out. Interviews took place over Attend Anywhere due to Covid-19 restrictions. An interview schedule (Appendix 2.8) was developed by the researcher in consultation with study supervisors, including a Consultant Clinical Psychologist working within the MND team. The schedule was also informed by the literature and included open questions with additional prompts. The schedule was used flexibly in line with IPA methodology (Smith et al., 2009). Interviews lasted between 40 minutes and 1 hour and 55 minutes. Interviews were audio-recorded after participants provided consent for this, before being transcribed verbatim. The researcher regularly checked in with participants in case of fatigue and/or distress. All potentially identifiable information was anonymised.

Analysis

Interpretative Phenomenological Analysis (IPA) was used to analyse data as it aims to explore participants' experiences, understandings, and perceptions, and assumes that participants seek to interpret their experiences into a form which is understandable to them. This method allowed research questions relating to people with MND's experiences of distress to be addressed.

The researcher utilised guidance from Smith et al. (2009) to analyse the data. Following verbatim transcription, interviews were listened to and read repeatedly by the researcher to become immersed in the data. Line-by-line analysis was then completed in order to note linguistic, descriptive, and conceptual themes. The next stage of analysis involved identifying themes, before identifying comparisons and contradictions of themes across participants. Throughout the analysis, the researcher moved back and forward through the transcript to reflect on what had previously been said, to interpret individual transcripts and all transcripts more fully as a whole. This was to ensure the double hermeneutic principles of IPA were incorporated into analysis as much as possible. Quotations were selected where they were thought to best capture the nature of the theme or where they provided particular insights. Samples of the transcripts were reviewed by one of the researchers to ensure that similar themes were identified (sample in Appendix 2.9).

Researcher Reflexivity

IPA places emphasis on researcher reflexivity, acknowledging that interpretations of data are tied to the researcher's ability to reflect and analyse. This is particularly important in this study as the researcher has no personal or professional experience of MND, therefore engaging in reflective practice allowed the researcher to reduce the impact of preconceptions on interpretations. The researcher is a female, trainee clinical psychologist with experience of qualitative methods (not IPA).

The researcher logged personal thoughts in a reflective journal to ensure reflexivity after each interview and during analysis (sample in Appendix 2.10). In line with IPA, the researcher approached the interviews openly while remaining mindful of potential bias.

Results

Sample Characteristics

Four participants lived with a partner and two participants lived alone with support from professional carers. Two participants had a slower progressing form of MND (30 years and 11 years since diagnosis). Mean time since diagnosis for the remaining participants was 1.3 years. Three participants were male (Andrew, Ben, and Tom) and three were female (Jane, Marion, and Sophie), with an average age of 73. One participant used a voice output device to communicate. Individual demographics are omitted due to risk of participant identification.

Qualitative Interviews

Six superordinate themes and ten subordinate themes emerged from the analysis, as summarised in Table 1.

1. Diagnosis related distress

The first superordinate theme details how participants made sense of their diagnosis. Two interrelated subthemes were identified.

***1.1* Distress when anticipating and receiving diagnosis.** All participants reported distress around waiting for and/or receiving a definitive diagnosis. Three participants stated that they had been aware of MND being a potential diagnosis. Tom, Anthony, and Ben reported feeling completely shocked, particularly Ben who had not known what MND was.

> *"…it sort of hits you like a brick wall. My god how did this happen, you know…I didn't know about MND. Didn't have a clue" Ben.*

Ben's use of *"hit me like a brick wall"* emphasises the sudden and significant negative impact of his diagnosis. Sophie and Jane reported feeling less initial shock as they had been expecting MND, with Jane feeling that the uncertainty of waiting for a confirmed diagnosis was almost as distressing as the diagnosis itself.

"I didn't know, until I got the diagnosis. Which was… you know, pretty hard in itself…but em, after all I'd been through it, um…it was worse probably, but, em…only slightly…" Jane

Participants all experienced fearing the worst, which was defined by suddenly confronting their own mortality. Fears also included how quickly they would deteriorate, and how they would cope with MND.

"…this is going to end my life prematurely, and between now and when that happens, I'm just going to get worse and worse and be able to do less and less. So…that was the distress." Sophie

Similar thoughts were described by Marion, *"…I was going to die, my life was finished."*, alongside the sense of there being no hope regarding treatment or cure. Both Andrew and Tom experienced the same initial distress. However, they both were eventually informed that their condition was progressing much slower than usual.

"Once I realised that it wasnae something…it wasnae going to happen tomorrow, I realised I could cope with it and I tried to act as normally as I possibly could." Andrew

There was a sense of them gaining back some control, and not experiencing the same sense of uncertainty as participants with a typical MND presentation.

1.2 Clinicians causing distress. Four participants experienced distress as a result of interactions with clinicians. Ben and Sophie had felt overwhelmed by the amount of information that they had received.

"Different teams, different medical people, were talking to me about different things…And I thought, yeah, alright, I know. And I think it was just one thing after another and it just was all stacking up and then it just all collapsed and it was just too much…" Sophie

The way that Sophie describes this experience demonstrates the impact of these stressors quickly building over a short time period. Ben reported being told "*virtually everything"* at the start which increased his distress and experience of uncertainty. There was a sense of being forced to confront the reality of the diagnosis before being ready to do so (“*yeah, alright, I know”*).

Tom and Jane experienced anger and frustration when dealing with clinicians. Jane described a GP misdiagnosing depression, despite her not experiencing low mood. Jane reported feeling frustration at how her own concerns were dismissed and feeling like she wanted vindication for this, stating “*I’ve always wanted to go back and say, I told you I wasn’t depressed”.’* Tom reported wishing to have his consultant struck off for the blunt manner in which his diagnosis was delivered to him.

> *“…a complete lack of empathy, he had no understanding of my potential agenda in a case like that, he was delivering, what was bound to be, life changing news, to a patient, and to a patient’s family, in an offhand way, without having anything to back it up, other than his…literally 5 minute examination of me.” Tom*

It was evident that Tom felt like he had not been involved in the diagnosis, describing feeling like a “*mute object”* and that the situation felt completely outside of his control.

Table 1.

Superordinate and Subordinate Themes

Superordinate	Subordinate
Diagnosis related distress	**Distress when anticipating and receiving diagnosis –** *"…I was going to die, my life was finished."*
	Clinicians causing distress – *"I thought it was absolutely dreadful doing that to anybody."*
Support influencing distress	**Valued support networks –** *"Everybody rallied round to help me."*
	Importance of specialist support – *"People are oblivious to the fact it exists."*
The distress of uncertainty	**Uncertainty as losing control –** *"You just never know."*
	Covid-19, MND and uncertainty – *"…nobody could come here, nobody could visit, it's all had to be masks."*
Loss of independence and roles	*"I'm very independent. I don't like depending on anybody."*
Coping with MND and distress	**Focusing on the positives –** *"You have to keep positive or you lose everything."*
	Maintaining normality and identity –*"I'm still me."*
Views of the MND Tool	**Reflecting experiences of MND –** *"It covers, most subjects to talk about, aye."*
	Enhancing person-centred care – "*You've got the option to say, yeah that affects me and to talk about it."*

2. Support influencing distress

The second superordinate theme considers the importance of both personal and professional support. There were two subthemes.

2.1 Valued Support Networks. All participants discussed the importance of support from family and friends. For Marion, Jane, and Sophie, this had been what they had been most grateful for, using variants of "*fortunate*" or *"lucky"*.

"I can't imagine how people that haven't got support around them…So I feel quite fortunate basically, I feel very lucky." Sophie

This sense of support being essential was reflected, with Ben reporting that his family "*do everything*" for him, Tom describing how "*everybody rallied roun*d" to help him and Jane stating that she "*couldn't have asked for more"* from her friends and family. Tom and Ben both described family members as being advocates for them.

"I still can fight my own corner, most of the time, but having a family member there helps me." Tom

This highlights Tom's desire to keep on doing as much as he can independently, while recognising that he will increasingly require support. Both Ben and Sophie described going through MND with their family, using *"we"* frequently, particularly when discussing coping with MND.

"We're all battling through it." Ben

Interestingly, it was also these two participants who discussed worrying about becoming a burden on their families and expressed guilt about having to depend more on their family.

"…it's only really him[husband] that I massively effect, um, so, so I do feel guilty about that." Sophie

Sophie repeated *"guilty*" when expressing not wanting to burden her adult children. Jane also alluded to this, reporting that her adult daughter had "*her own things to contend with*". Ben reported being concerned about *"all of the extra work*" that he feels that he is passing onto his family). Andrew also described the *"frustration*" of having to depend on his wife to carry out specific tasks for him. This highlights that although support is seen as positive and essential, it can also elicit difficult emotions.

2.2 Importance of Specialist Support. Specialist support, such as the MND team and MND Scotland was seen as person centred, knowledgeable, helpful, and responsive to people's needs. Andrew reported having a positive experience of all services involved in his care, but was particularly positive about the MND team.

> *"Anything I've asked them for they've...managed to provide…what I needed. There's someone on the end of the phone if I need them, eh…it makes me quite…quite happy, aye." Andrew*

It appeared that having support available helped him to maintain some control. Tom echoed this experience of the MND team and highlighted their collaborative approach as being particularly beneficial *("…they participate with me in any discussions and I'm included in things")*. Tom discussed the uncomfortable need to gradually pass control to professionals, and reported that still feeling involved made this easier. Ben also emphasised his appreciation of collaborative care, describing appointments at the MND clinic as being *"50/50*". Marion acknowledged that this *was "…usually better, more successful, than other interactions"* , and echoed Andrew's sense of control in knowing that there *is "…support for most situations".* Ben highlighted the role of reliable support in helping to reduce worry and distress.

> *"The only positive thing I can say is to do with the MND team…I'm not worried about anything at all and it's all down to those people."* Ben

Andrew and Tom, who both have slower progressing MND, had very strong views on the general lack of awareness of MND particularly in comparison to other conditions.

> *"…cause it's not cancer…it's motor neurone disease, and people don't know what to do about that. Most people are not aware of what that is, even, and…they don't regard it as being, any kind of a priority…So you just get dealt with in the queue." Tom*

Tom emphasised the lack of awareness specifically being around timing of support. Jane reported non-specialist services as not providing person-centred care and having a lack of awareness of MND:

> *"… you know, they don't understand about MND or…I'm just a, you know, body at the bed…." Jane*

Jane and Sophie reported that specialist supports were much more responsive when providing adaptations but that generally their provision felt like *"…reacting rather than anticipating"*. Jane acknowledged the possibility of patients feeling *"despondent"* if given adaptations too quickly. Sophie contrasted specialist and non-specialist services to emphasise the difference in responsiveness:

> *"…I can just ring [MND nurse] and she sets the wheels in motion and things happen… but [non-specialist service]…that's been a long time, when I really needed it more then…" Sophie.*

3. The distress of uncertainty

The third superordinate theme highlights the inherent distress of uncertainty in MND. Two interrelated subthemes were identified.

3.1 Uncertainty as losing control. The difficulty of living with uncertainty as part of MND was reported by all participants with the exception of Tom (*"…I don't dread it at all."*) who has slower progressing MND, which appeared to alleviate uncertainty. In contrast Andrew, did find the experience of uncertainty around deterioration distressing.

> *"The uncertainty of when it is going to happen is uh…the most, most sort of, concerning thing. I mean, it might be a while yet, I don't know when it will be, and that's…that's the thing." Andrew*

The way Andrew explains this demonstrates the back and forward nature of his thoughts. All five participants struggling with uncertainty described variants of *"you just never know*" and *"you don't know what's coming*". Marion highlighted that this uncertainty is pervasive: *"…it is difficult to live with daily*." Ben and Jane both emphasised that changes come quickly and unexpectedly. For Ben, physical changes felt constant and unpredictable.

> *"You know… totally unexpected things happen. Like my speech, my mobility, my breathing, um… loss of power in my hands and arms…legs, you know, it's just progressing, one thing after another" Ben.*

This again highlights that uncertainty is a constant presence in people's lives. Jane emphasised that even when you aware of what changes you are likely to experience, you cannot predict the impact these will have on you or how you will cope when these changes happen.

> *"I knew I would get worse…but, um, it's only really when you're going through it you realise, at…at each stage how it affects what you can do. I'm just, at the stage of, how will I cope if I'm any worse." Jane*

For all participants, there was a sense of this uncertainty resulting in a sense of feeling like nothing was in their control. Sophie in particular emphasised the difficulties of not being able to make plans due to how quickly deterioration can occur.

> *"But you can't move forward, you can't make plans, you can't make plans at all. Um, yeah, it's difficult not being able to plan anything, so that's hard, that's distressing." Sophie*

Sophie also struggled with the loss of the plans she had for her future. Uncertainty is one of the main areas of distress that people with MND have to cope with as it is an inherent part of the progressive nature of the condition.

3.2 Covid-19, MND and uncertainty. Three participants (Ben, Jane, and Sophie) had received their diagnosis during the coronavirus pandemic. Covid-19 related restrictions and lockdowns were perceived as both exacerbating MND related uncertainty and as masking changes. These participants viewed the experiences of MND and going through covid as being intertwined and hard to distinguish between.

"Yeah, it all came at the same time, so I can't really…differentiate between getting MND and going through covid, it's all came at the one time. So going through both at the one time, that's all we can do." Ben

Ben used "we" to highlight that he got through both with his family. Having a common experience of uncertainty with others appeared to be helpful for Sophie.

"…they were limited by covid restrictions and I was limited by MND and covid, but, you know, they were restricted and now all of a sudden everybody's out doing stuff…" Sophie

For Sophie, this shared uncertainty of Covid-19 had eased the uncertainty around her MND, as others had not noticed the impact of her being able to do less. However, she also acknowledged that Covid-19 made it even more difficult to make plans, and how she may have spent her time differently if she had been diagnosed at another time. Jane reflected on what she could do before lockdown compared to now, and therefore restriction emphasised changes for her. She also noted similarities between lockdown and MND both making days difficult to distinguish between.

"With lockdown it's hard to tell which day of the week it is (laughs). Every day is much the same, it's the same routine and I have to stick to a routine because the carers come… according to a schedule." Jane

4. Loss of independence and roles

All participants reported finding loss of independence as a result of MND distressing. Andrew's fear of uncertainty centred around when he would need a wheelchair, and this appeared to be his biggest source of distress due to a fear of losing his independence.

"I want my independence; I don't want to be dependent on other people pushing me about on a wheelchair." Andrew

The fear of losing mobility was understandably linked with a fear of losing independence. Tom described how he could still walk, but weakness in his upper limbs had left him with a fear of falling.

"I'm afraid of falling, I know that if I do fall, if I fall that way, that will be the end of me…" Tom

This portrays a sense of Tom feeling that if he falls, he will lose mobility and be unable to do anything independently. *"…the end of me"* demonstrates how dreaded an outcome this would be for him. Andrew reported finding it distressing and frustrating to have to rely on family members to perform tasks he would normally do himself, *"I get very frustrated cause I cannae do that."* Andrew viewed himself as someone who had always been independent, therefore gradually losing this ability was the most distressing consequence of MND. Similarly, Ben found this loss of control and increasing need to depend on his family to be distressing.

"… I used to do virtually everything for them, and now the tables turned completely and they're having to do everything for me. So that is hard to get your head round and to accept. Yeah." Ben

This description demonstrates how much this was experienced as a complete role change for Ben, which has felt difficult to comprehend. There was a sense that for most participants, being able to function independently was inextricably linked with their identity. Sophie and Jane both experienced a loss of independence.

"…I was self-sufficient, with help, and now I'm not." Sophie

Sophie's description emphasises that she was previously able to do everything herself, and how this has quickly declined. Sophie and Jane also found it difficult that their role had shifted from supporters to being supported, particularly as grandparents not being able to support their children:

"I used to be able to go and help my daughter at times…or other members of the family I used to try and help but…I can't do all that now..." Jane

There was a sense of guilt for not being able to physically provide support for family members, as well as a loss of not being able to participate in family roles as expected. For Marion, her loss of independence came from losing her ability to verbally communicate. She described how this impacted her ability to be involved in activities independently.

"You need to verbally communicate. You feel vulnerable in certain situations…. And people relate to you in a different way, not as an equal. Probably because I have no voice." Marion

Marion's description suggests that losing her voice has resulted in losing her ability to be involved and to feel in control in social situations, therefore feeling more dependent on others. Marion also described how fatigue results in a frustrating disparity between what she wants to do and what she has the energy to do independently.

5. Coping with MND and distress

The fifth superordinate theme describes how people cope with MND and distress. Two interrelated subthemes were identified.

5.1 Focusing on the positives. A coping strategy for all participants was to focus on the positives in the past or present rather than on the difficulties of MND or the uncertainty of the future. There was a sense of this being the only choice that people had and that this was a conscious effort to choose to focus on the small positives.

"I always try and think…think positive and I try not to think about…about the unpleasant thing. But it's not that I… put my head in the sand, you know, you can't forget them, but you just have to, concentrate on the good things, you know…getting outside to see the daffodils... " Jane

Jane emphasises that it is not about avoidance, as this is impossible. This demonstrates that people still feel distressed, but that concentrating on the positives is more helpful than thinking about what they have lost. There was a sense of people trying to make the most of the time that they had left rather than thinking about what they no longer have.

"There are many losses, but you have to look for new positives and be grateful." Marion

Andrew and Ben emphasised that this is a conscious effort (*"I don't let things get me down."* Andrew) and simultaneously the only choice to avoid further distress (*"…there's nothing I can do except accept it and try to carry on...."* Ben). Jane, Sophie, and Tom all reported finding it helpful to reflect back on the positive experiences they had throughout their lives.

> *"…despite everything, we've been quite lucky, well, whether you call it lucky, we're really, you know, happy with what we've managed to do so far." Sophie*

This portrays that people are able to manage some of the distress they are experiencing in the present by focusing on pleasant memories from their past (*"…remember that you had all these good times."* Jane), creating a sense of being satisfied with the life they have lived (*"I've done all the things that I wanted to do."* Tom).

5.2 Maintaining Normality and Identity. Accepting MND for people meant accepting but not giving in to the diagnosis. Participants reported coping by trying to do as much as they could for as long as possible.

> *"… if I can fight it by…trying, trying to talk, walk about, do things with my hands and arms, if I can do it, I'll carry on, until I can't do it anymore." Ben*

Maintaining normality seemed to give people a sense of control by not giving in. Sophie emphasised the importance of still planning some things and trying to adapt to ongoing changes, but how this can be difficult when their adaptations stop working.

> *"…you think you've got something sorted out, and it all comes crashing down, and then you end up quite low. Until you get up the next morning and you think, right, we'll have to find something else to do. And you attack it and get started again, you know, trying to find ways to keep doing things." Sophie*

This sense of not giving in appeared to be what motivated people to stay in the present, and to remain positive, involved and in control. Andrew and Tom had both made adaptations to their homes themselves, anticipating what problems they would be facing in the future in an attempt to maintain normality and a sense of control for as long as possible.

“Being proactive obviously doesn’t change things or the likely outcome of things, but it enables you to go on staying in control as much as you can for as long as you can.” Tom

Maintaining normality allowed participants to feel like they were still in control in the face of uncertainty, and part of this was maintaining their own identity. All participants reported that although MND has impacted on them physically, MND has not impacted on their identity (*“I’m still me”* Jane). This appeared to be a source of strength that people drew upon, with participants describing themselves as *“…quite a confident person. I’m quite positive”* (Andrew), *“a coper and problem solver*” (Sophie), *“a very proactive person”* (Tom) and *“I’m not one for just packing up”* (Ben).

6. Views of the MND Tool

The final superordinate theme portrays participant’s views of the MND Distress Thermometer (DT) and Concerns Checklist (CC). There were two interrelated subthemes.

6.1 Reflecting experiences of MND. All participants described the DT/CC tool as useful or helpful, and felt it covered relevant areas. All participants stated that they did not feel that was anything missing, with three participants describing the tool as comprehensive.

“I think it’s very useful, to have a structured approach.” Tom

Three participants commented that they felt that their own concerns fit into the checklist, although Sophie stated that she could not pick concerns to prioritise as they were all being dealt with. Jane reported that the categories were a helpful memory prompt for concerns that may not affect people all the time, but found the idea of rating her distress on a scale to be difficult.

“It’s hard to…categorise things, things such as feelings, but…it’s very hard to um, quantify.” Jane

Both Sophie and Ben reported worrying about areas that were not affecting them now potentially impacting on them in the future. However, this did not appear to be overly distressing; Sophie reflected that the list could remind people of what they can do, and that she was already aware of the possible changes. Ben felt that he would be able to cope with any changes that came his way, as he had been doing.

"So you've got a few things down there which make you wonder. It really could make you worry. As I've said, anything that's for me, will come this way, and I'll just accept it and carry on the best I can." – Ben

6.2 Enhancing person-centred care. All participants agreed that the tool would be useful to guide a discussion with their HCPs. All participants referred to the importance of the tool being used to ensure people are able to discuss their concerns. Four participants felt that they would not need to use the tool to raise concerns, but felt that the tool would be particularly helpful for giving control and choice to people who find it difficult to raise issues with HCPs.

"I can still do that [speak up for himself]…But many patients won't be able to do those things, and obviously to have something structured for them is something that could help them to do that." Tom

Marion and Tom both felt that a potential issue with the tool could be HCPs being unwilling to use the tool collaboratively. It is worth noting that these participants had experienced some HCPs as being unempathetic or as very directive.

"I think the tool would help if the clinician was open to using it with you." Marion

Discussion

The current study explored the lived experience of distress in the context of MND. Additionally, MND patients' views on using the Distress Thermometer and Concerns Checklist were examined. Following IPA analysis, six superordinate themes emerged: i) diagnosis related distress, ii) support influencing distress, iii) the distress of uncertainty, iv) loss of independence and roles, v) coping with MND and distress and vi) views of the MND tool.

Consistent with existing research, the current study highlights diagnosis of MND as a point of distress, even when people desire a definitive explanation for symptoms (Remm et al., 2019; Aoun et al., 2016). A study exploring experiences of MND diagnosis found that factors which mediated distress were HCPs who treated patients as individuals and support from family and friends (Remm et al., 2019), reflecting findings in the current study. Aoun et al. (2016) highlighted neurologists specifically could improve patient experiences by demonstrating more empathy, increasing consultation times, decreasing follow-up periods, and sharing more information. In the current study, participants reported experiencing empathy and care from specialist services but not from general health and social care services.

The importance of support from family and friends in alleviating distress was highlighted by participants. Research within palliative care has suggested that this support is a protective factor, the absence of which may identify patients at risk of struggling to cope (Milberg et al., 2014). Additionally, Leigh et al. (2021) found that MND patients with higher levels of social support had lower levels of psychological distress. Some participants were concerned about the burden their family caregivers may experience. However, qualitative research has found that family caregivers can also experience acceptance, gratitude, hope and happiness (Tan et al., 2020). This study highlighted the need for caregivers to be supported, and how HCPs can help by involving them in decision making and by communicating empathetically.

Participants felt there was a general lack of awareness of MND. Lack of knowledge from professionals has been identified as one barrier to diagnosis (Remm et al., 2019). Lack of awareness from HCPs can reduce the efficacy of advanced care planning, impacting on interventions patients receive (Murray & Butow, 2016). Participants in one qualitative study (Weeks et al., 2019) felt there was a lack of awareness, both societally and among non-MND clinicians, and similarly contrasted this with increased awareness of cancer. Participants reported that this led to inadequate provision of information and support, as well as feelings of isolation.

The current study highlighted the need for person centred care, beginning while people are seeking a diagnosis. Paynter et al. (2020) described how decision making for some was seen as being limited to accepting or rejecting recommendations and having a choice around *when* to accept interventions rather than *if*. This feeling of not having control could be a barrier to shared decision making in MND. Person centred support can help people to maintain control over some aspects of their lives in the face of uncertainty, by remaining involved in their care (Warnock et al., 2017).

Results reflect findings that the experience of uncertainty is pervasive and begins while seeking a diagnosis of MND (Remm et al., 2019). A study exploring uncertainty in MND (Harris et al., 2018) reflected themes in the current study around how maintaining control in the face of uncertainty through being involved in health care interventions can result in a further loss of control (e.g., need for professional carers). It also stressed how coping is easier for those who can accept the disease.

The current study discussed the challenges that Covid-19 brought in terms of exacerbating uncertainty and exaggerating difficulties. Glasmacher et al. (2021) outlined the impact of the pandemic on MND patients, including anxiety around exposure, difficulties receiving support, loneliness, and fear of being denied treatment. This demonstrates the wider impact that Covid-19 has had on people with MND.

Loss of independence and roles for participants in the current study were identified as sources of distress. Soundy & Condon (2015) discussed changes to roles and independent living as major losses, while Munan et al. (2021) connected these two experiences; people who experienced role reversal with their partner experienced a loss of independence. They also reported that role changes can result in a loss of intimacy with disease progression. Sakellariou et al. (2013) highlighted that people with MND have no control over the disease, but they can be supported to maintain control over their care and to feel as independent as possible.

Participants in the current study reported coping with MND in similar ways to participants in Remm et al. (2019); by regaining control, and maintaining normality and identity for as long as possible, strategies also found in other studies (Soundy & Condon, 2015; Sakellariou et al., 2013). Some patients reported finding these methods of coping difficult in the face of feeling overwhelmed with their diagnosis. Ways of coping reported in the present study fit with the idea of an internal health locus of control e.g., by focusing on areas of their disease that they do have control over as opposed to aspects which are outside of their control (external locus of control). In cancer patients, an internal locus of control is associated with better coping strategies (Goldzweig et al., 2016). In a study which examined therapeutic approaches for people with MND (Rabbitte et al., 2015), most focused on supporting individuals to maintain an internal locus of control. It is likely to be more difficult to focus on an internal locus of control in a MND, where there is no hope for cure, but it may be beneficial for patients if they are supported to do so.

Participants in the current study felt that the MND Distress Thermometer and Concerns Checklist were comprehensive tools which allowed for the facilitation of collaborative discussions. Some participants felt that clinicians may be resistant to using this, which reflected their own experiences of care not being person centred. Unfortunately, participants had not had the opportunity to use these tools in practice, although points raised reflected some of the experiences of cancer patients who had used the DT and CC. Biddle et al. (2016) found that patients found the tools useful in identifying and validating distress, improving holistic care, improving relationships with HCPs and for increasing the potential for concerns to be addressed.

Participant responses indicated that they may have specific concerns (e.g., about requiring a specific adaptation soon) without being distressed. However, participants discussed how concerns could turn into sources of distress if they were not addressed. Experiences of distress appeared to fit Brennan et al.'s (2012) definition, encompassing a range of factors which can cause distress. However, it is worth noting that Biddle et al. (2016) found that patients did not always report distress, as they defined this with regards to significant mental health difficulties rather than concerns impacting on their life and/or wellbeing. Sousa et al. (2021) discussed the importance of informing participants using the DT about the definition of distress.

Strengths and Limitations

These findings represent the personal experiences of a small, homogenous group of people with MND, which limits their generalisability. However, generalisability is not the aim of IPA, as this methodology seeks to gain an in-depth understanding of experiences. Samples of the transcripts were reviewed by one of the supervisors of this research to increase reliability. If resources had allowed, further measures could have been taken to limit subjectivity, such as researchers independently coding themes and reviewing themes with participants. The main strength of the study is the addition to the literature of a rich, descriptive phenomenology of distress in MND, which can inform practice and research.

Recruitment proved difficult, due to demands placed on the MND team and the additional demands of the Covid-19 pandemic. MND clinicians acknowledged feeling apprehensive about approaching patients as they did not want to place another demand on people. This may have resulted in a bias in recruitment, where participants are people who have been more able to cope with the distress of MND. Participants may have felt that they had to be positive about the MND team, although they were reassured that the study was independent from the clinic prior to interview. Originally this study would have sought to explore their personal experiences using the tool, but unfortunately this has not been used as part of clinical appointments due to the majority of appointments taking place remotely. This may have resulted in participants being unclear about the use of the tool. IPA would have been appropriate to explore participants ʾlived experiences of using the tool, and it was decided that it would still be useful to interview participants about how they felt the tool related to their experiences of distress. However, as participants did not experience using the tool it may have been more appropriate to investigate these views using a different method of analysis (e.g., thematic analysis) as using IPA for this aspect of the study did not provide the same rich, descriptive data as provided by the rest of the interview questions.

Participants received support from the same specialist MND team, therefore many experiences of HCPs will be from the same service. However, local services would have covered a wider range of HCPs. Participants were of similar ages. People diagnosed with MND at a younger age may have had different experience of distress. The experience of distress in MND may differ depending on disease progression. Two participants had a slower progressing MND, which did appear to have an impact on their experiences of distress, although there were similarities to other participants' experiences too.

For one participant communication difficulties made an in-depth exploration of experiences difficult. This participant met inclusion criteria as they could verbally communicate using a voice output device. The use of alternate communication methods was out with the scope of this study, but would be a valuable addition to future research.

Clinical Implications

The findings of this study highlight the importance of person-centred care, which can help patients to maintain independence, which may enhance their ability to use helpful coping strategies. Increasing awareness of MND in non-specialist services may help HCPs to provide better care.

Timely and accurate information about diagnosis, prognosis and treatment options can help patients to feel more empowered. Sakellariou et al. (2013) highlighted that it can be difficult to achieve a balance between both the amount and timing of information, with people preferring to deal with problems as they come up rather than too far in advance. Information should be easily understood and individualised to the patient as much as possible to support this. This study highlighted the importance of family support, suggesting that there should be a collaborative treatment plan between HCPs, the patient, and their families.

The nature of MND makes decision making challenging. Utilising tools such as the MND DT and CC can help patients to be involved in their care, maintaining a sense of control and independence which can help mediate the impact of uncertainty. These tools can enhance patient centred care by supporting discussions between HCPs, patients, and their families. Research on tools to involve patients in decision making indicate that their use can increase engagement, improve communication between patients and HCPs and reduce unnecessary treatment (Stacey et al., 2017). These tools should be subject to ongoing evaluation with the views of patients being incorporated.

Services using the MND DT and CC should make this available online. This would allow the tools to be easily updated with new information, and allow patients to go through this with clinicians during phone or video appointments. Training should be provided to HCPs around how to use the tool in a collaborative manner and to ensure that they feel comfortable discussing distress. There should be clear pathways for clinicians around where to refer patients to for support if required. Tools should be administered frequently due to the rapidly changing nature of MND.

Although further research is required around psychological interventions in MND, coping strategies described in the current study suggest that approaches such as Acceptance and Commitment Therapy (ACT) or mindfulness may be beneficial. Weeks et al. (2019) highlight aspects of ACT which may be beneficial to people with MND, such as a focus on increasing valued behaviour, and differentiating between what can and cannot be controlled.

Future Directions

Increased understanding of information provision could allow HCPs to deliver more person-centred care. Future research could focus on participants' experience of distress, care, and information provision longitudinally to see how these experiences change over time. Conducting interviews using different communication aids would assist with ensuring inclusion.

Future research should focus on how both patients and clinicians experience using the MND DT and CC in order to identify benefits and barriers associated with its use. Biddle et al. (2016) found that clinicians and patients identified different barriers to using these tools in an oncology setting. Further research may want to investigate the validity of the MND DT or internal consistency of the MND CC.

Clinicians may be wary about approaching patients about research participation. However, including people in research may enhance their feelings of being involved and autonomous, which would link in with identified helpful coping strategies in the current study. Research could investigate how patients with MND experience this.

Conclusions

This study sought to explore experiences of distress in patients with MND. Findings highlighted the pervasive nature of uncertainty, loss of roles and independence in MND and the impact that support can have on distress. Participants in the current study also demonstrated extraordinary resilience, reporting coping by focusing on positives, and on maintaining normality. Findings highlight the need for person centred care, which can be improved by the use of tools such as the MND DT and CC.

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Appendices

Appendix 1.1 Author Guidelines for Submission to the International Journal of Qualitative Studies on Health and Well-being

Aims and scope

QHW provides a forum for the exchange of data, knowledge, theoretical framework and methods on health and well-being. The journal aims to further the development and understanding of qualitative research by using rigorous qualitative methodology of significance for issues related to human health and well-being.

International Journal of Qualitative Studies on Health and Well-being publishes empirical research, with a national and/or international focus, as well as articles on:

- Qualitative research approaches
- Qualitatively-driven mixed-method designs
- Qualitative methodological development
- Meta-synthesis
- Theoretical and philosophical issues related to qualitative research and health and well-being

Please note that this journal only publishes manuscripts in English.

International Journal of Qualitative Studies on Health and Well-being accepts the following types of article: Empirical Studies, Review Articles, Philosophical Papers.

Preparing Your Paper

Structure

Your paper should be compiled in the following order: title page; abstract; keywords; main text introduction, materials and methods, results, discussion; acknowledgments; declaration of interest statement; references; appendices (as appropriate); table(s) with caption(s) (on individual pages); figures; figure captions (as a list).

Word Limits

Please include a word count for your paper. There are no word limits for papers in this journal.

Format-Free Submission

Authors may submit their paper in any scholarly format or layout. Manuscripts may be supplied as single or multiple files. These can be Word, rich text format (rtf), open document format (odt), or PDF files. Figures and tables can be placed within the text or submitted as separate documents. Figures should be of sufficient resolution to enable refereeing.

- There are no strict formatting requirements, but all manuscripts must contain the essential elements needed to evaluate a manuscript: abstract, author affiliation, figures, tables, funder information, and references. Further details may be requested upon acceptance.
- References can be in any style or format, so long as a consistent scholarly citation format is applied. Author name(s), journal or book title, article or chapter title, year of publication, volume and issue (where appropriate) and page numbers are essential. All bibliographic entries must contain a corresponding in-text citation. The addition of DOI (Digital Object Identifier) numbers is recommended but not essential.
- The journal reference style will be applied to the paper post-acceptance by Taylor & Francis.
- Spelling can be US or UK English so long as usage is consistent.

Checklist

1. **Author details.** All authors of a manuscript should include their full name and affiliation on the cover page of the manuscript. Where available, please also include ORCiDs and social media handles (Facebook, Twitter or LinkedIn). One author will need to be identified as the corresponding author, with their email address normally displayed in the article PDF (depending on the journal) and the online article. Authors' affiliations are the affiliations where the research was conducted. If any of the named co-authors moves affiliation during the peer-review process, the new affiliation can be given as a footnote. Please note that no changes to affiliation can be made after your paper is accepted. Read more on authorship.
2. Should contain a structured abstract of 200 words.

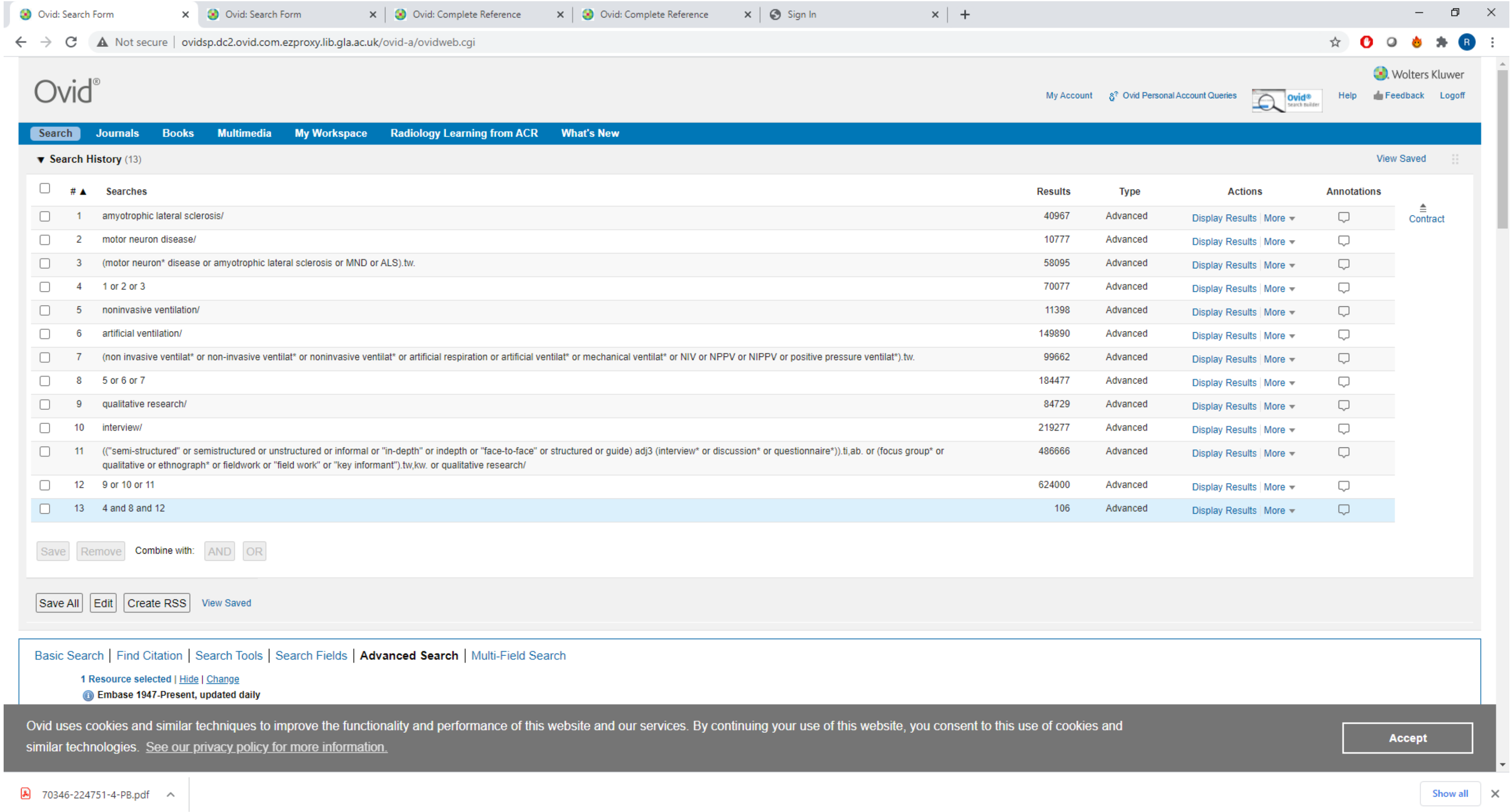
   A structured abstract should follow the structure: Purpose, Methods, Results, and Conclusions.

See the following link for further details:

https://www.tandfonline.com/action/authorSubmission?show=instructions&journalCode=zqhw20

Appendix 1.2 Search Terms and Results by Database

Embase	
1	Amyotrophic Lateral Sclerosis/ [SH]
2	Motor Neuron Disease/ [SH]
3	(motor neuron* disease or amyotrophic lateral sclerosis or MND or ALS).tw [Title or abstract]
4 [MND Component]	1 OR 2 OR 3
5	Noninvasive ventilation/ [SH]
6	Artificial ventilation/ [SH]
7	(non invasive ventilat* OR non-invasive ventilat* OR noninvasive ventilat* OR artificial respiration OR artificial ventilat* OR mechanical ventilat* OR NIV OR NPPV OR NIPPV OR positive pressure ventilat*).tw (title or abstract)
8 [NIV component]	5 or 6 or 7
9	Qualitative research/
10	Interview/
11[Qualitative filter]	((("semi-structured" or semistructured or unstructured or informal or "in-depth" or indepth or "face-to-face" or structured or guide) adj3 (interview* or discussion* or questionnaire*))).ti,ab. or (focus group* or qualitative or ethnograph* or fieldwork or "field work" or "key informant").tw,kw or qualitative research/
12 [qualitative component]	9 OR 10 OR 11
10 [whole search]	4 AND 8 AND 12



Search History (13)

# ▲	Searches	Results	Type	Actions	Annotations
1	amyotrophic lateral sclerosis/	40967	Advanced	Display Results | More	
2	motor neuron disease/	10777	Advanced	Display Results | More	
3	(motor neuron* disease or amyotrophic lateral sclerosis or MND or ALS).tw.	58095	Advanced	Display Results | More	
4	1 or 2 or 3	70077	Advanced	Display Results | More	
5	noninvasive ventilation/	11398	Advanced	Display Results | More	
6	artificial ventilation/	149890	Advanced	Display Results | More	
7	(non invasive ventilat* or non-invasive ventilat* or noninvasive ventilat* or artificial respiration or artificial ventilat* or mechanical ventilat* or NIV or NPPV or NIPPV or positive pressure ventilat*).tw.	99662	Advanced	Display Results | More	
8	5 or 6 or 7	184477	Advanced	Display Results | More	
9	qualitative research/	84729	Advanced	Display Results | More	
10	interview/	219277	Advanced	Display Results | More	
11	(("semi-structured" or semistructured or unstructured or informal or "in-depth" or indepth or "face-to-face" or structured or guide) adj3 (interview* or discussion* or questionnaire*)).ti,ab. or (focus group* or qualitative or ethnograph* or fieldwork or "field work" or "key informant").tw,kw. or qualitative research/	486666	Advanced	Display Results | More	
12	9 or 10 or 11	624000	Advanced	Display Results | More	
13	4 and 8 and 12	106	Advanced	Display Results | More	

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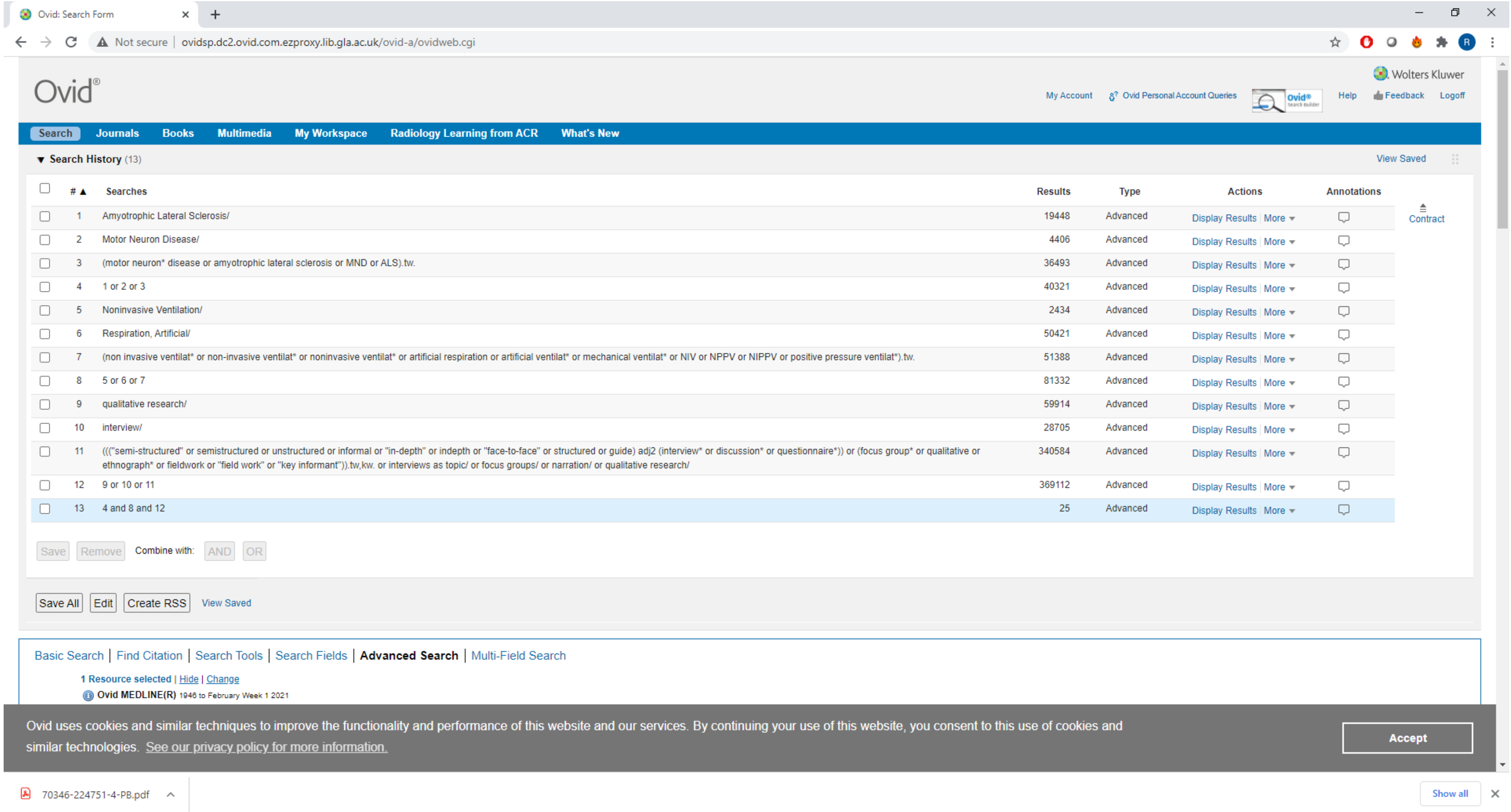
Medline	
1	Amyotrophic Lateral Sclerosis/ [SH]
2	Motor Neuron Disease/ [SH]
3	(motor neuron* disease or amyotrophic lateral sclerosis or MND or ALS).tw [Title or abstract]
4 [MND Component]	1 OR 2 OR 3
5	Noninvasive ventilation/ [SH]
6	Respiration, Artificial/ [SH]
7	(non invasive ventilat* OR non-invasive ventilat* OR noninvasive ventilat* OR artificial respiration OR artificial ventilat* OR mechanical ventilat* OR NIV OR NPPV OR NIPPV OR positive pressure ventilat*).tw (title or abstract)
8 [NIV component]	5 or 6 or 7
9	Qualitative research/
10	Interview/
11 [Qualitative filter]	((("semi-structured" or semistructured or unstructured or informal or "in-depth" or indepth or "face-to-face" or structured or guide) adj2 (interview* or discussion* or questionnaire*))).tw,kw or (focus group* or qualitative or ethnograph* or fieldwork or "field work" or "key informant").tw,kw or interviews as topic/ or focus groups/ or narration/ or qualitative research/
12 [qualitative component]	9 OR 10 OR 11
10 [whole search]	4 AND 8 AND 12



▼ Search History (13)

☐	# ▲	Searches	Results	Type	Actions	Annotations
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☐	2	Motor Neuron Disease/	4406	Advanced	Display Results | More ▾	🗨
☐	3	(motor neuron* disease or amyotrophic lateral sclerosis or MND or ALS).tw.	36493	Advanced	Display Results | More ▾	🗨
☐	4	1 or 2 or 3	40321	Advanced	Display Results | More ▾	🗨
☐	5	Noninvasive Ventilation/	2434	Advanced	Display Results | More ▾	🗨
☐	6	Respiration, Artificial/	50421	Advanced	Display Results | More ▾	🗨
☐	7	(non invasive ventilat* or non-invasive ventilat* or noninvasive ventilat* or artificial respiration or artificial ventilat* or mechanical ventilat* or NIV or NPPV or NIPPV or positive pressure ventilat*).tw.	51388	Advanced	Display Results | More ▾	🗨
☐	8	5 or 6 or 7	81332	Advanced	Display Results | More ▾	🗨
☐	9	qualitative research/	59914	Advanced	Display Results | More ▾	🗨
☐	10	interview/	28705	Advanced	Display Results | More ▾	🗨
☐	11	((("semi-structured" or semistructured or unstructured or informal or "in-depth" or indepth or "face-to-face" or structured or guide) adj2 (interview* or discussion* or questionnaire*)) or (focus group* or qualitative or ethnograph* or fieldwork or "field work" or "key informant")).tw,kw. or interviews as topic/ or focus groups/ or narration/ or qualitative research/	340584	Advanced	Display Results | More ▾	🗨
☐	12	9 or 10 or 11	369112	Advanced	Display Results | More ▾	🗨
☐	13	4 and 8 and 12	25	Advanced	Display Results | More ▾	🗨

1 Resource selected | Hide | Change

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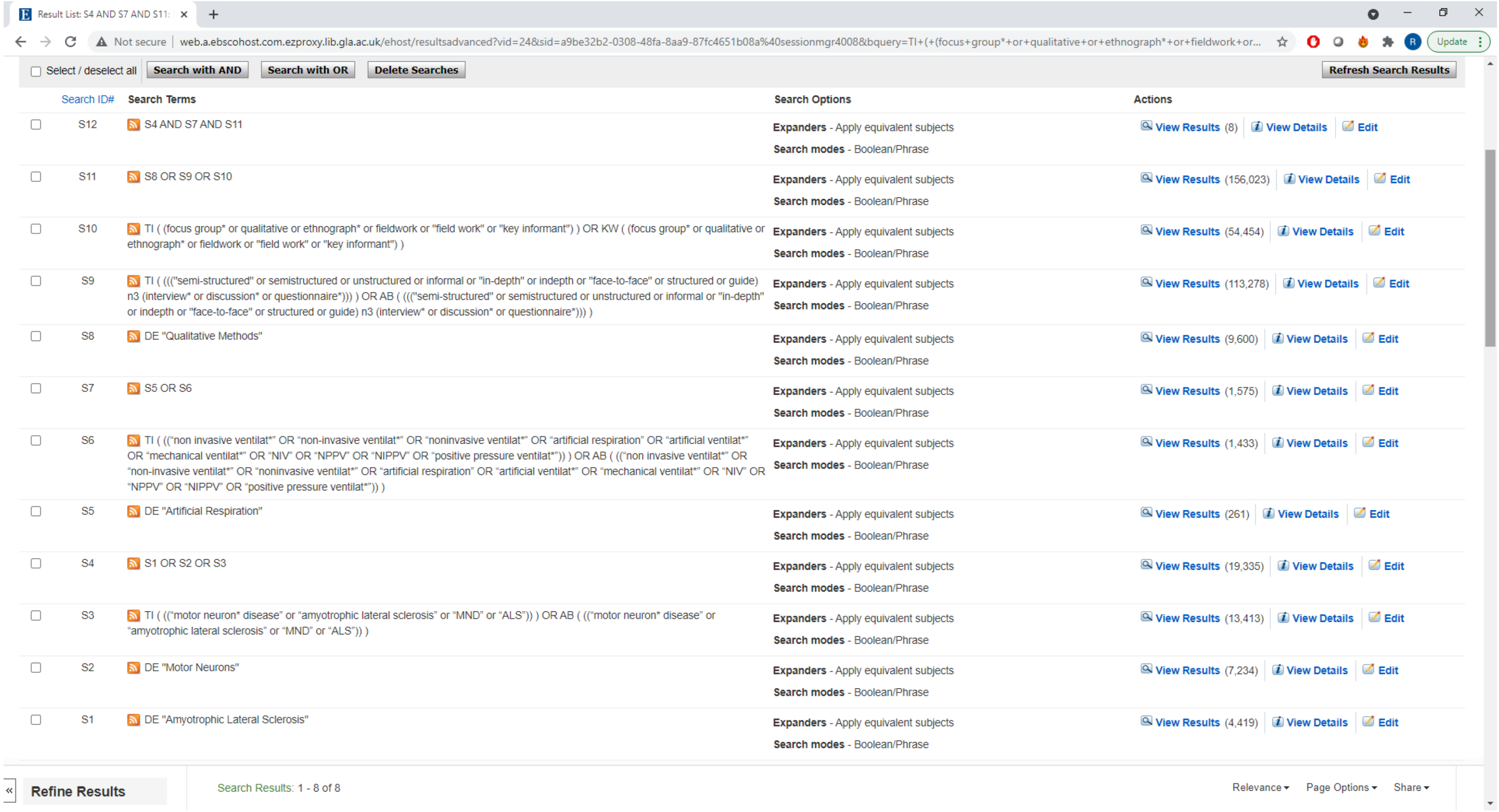
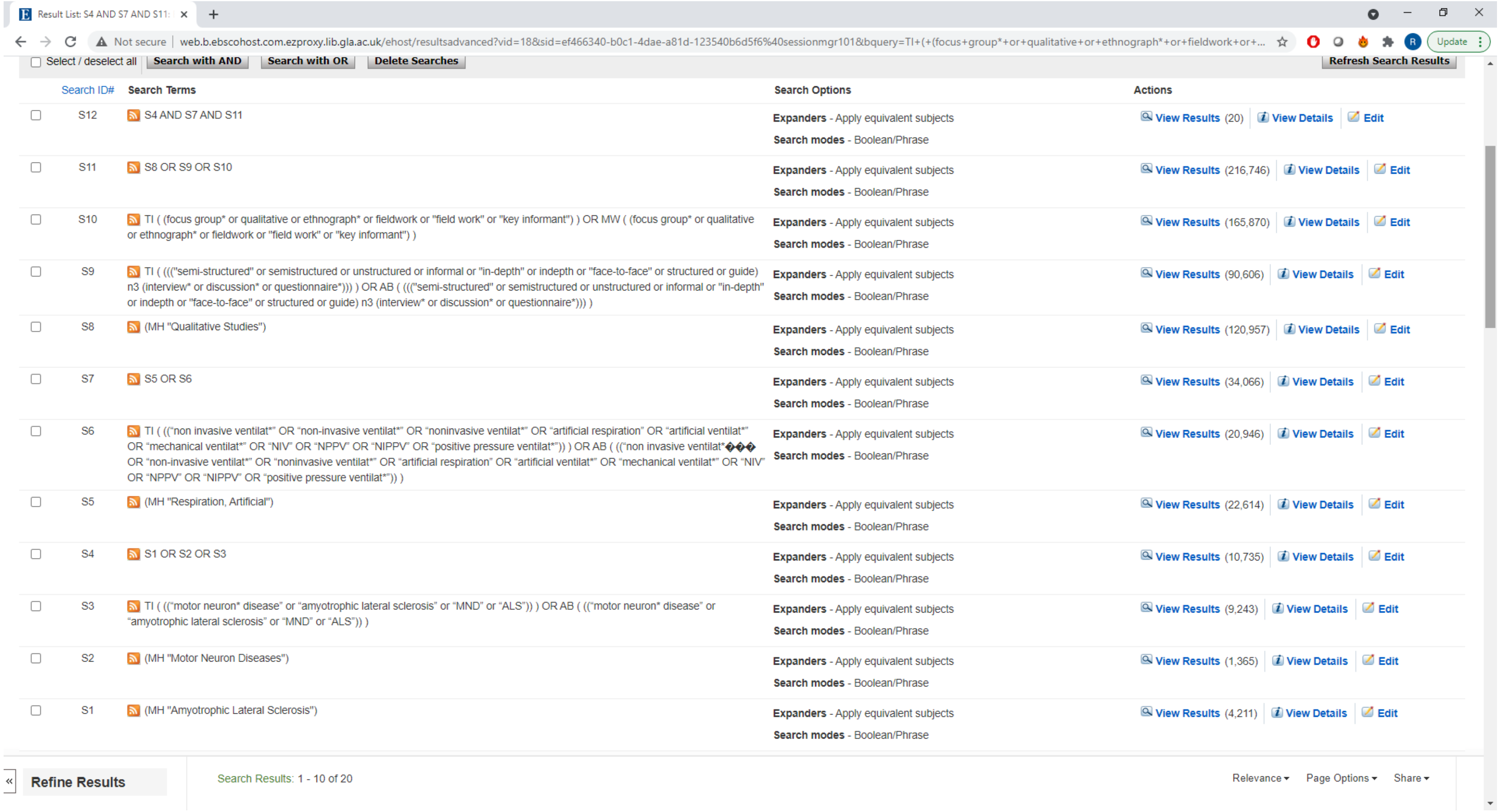
PsycInfo	
1	DE “Amyotrophic Lateral Sclerosis” [SH]
2	DE “Motor Neurons” [SH]
3	TI ((“motor neuron* disease” or “amyotrophic lateral sclerosis” or “MND” or “ALS”)) OR AB ((“motor neuron* disease” or “amyotrophic lateral sclerosis” or “MND” or “ALS”))
4 [MND Component]	1 OR 2 OR 3
5	DE “Artificial Respiration” [SH]
6	TI ((“non invasive ventilat*” OR “non-invasive ventilat*” OR “noninvasive ventilat*” OR “artificial respiration” OR “artificial ventilat*” OR “mechanical ventilat*” OR “NIV” OR “NPPV” OR “NIPPV” OR “positive pressure ventilat*”)) OR AB ((“non invasive ventilat*” OR “non-invasive ventilat*” OR “noninvasive ventilat*” OR “artificial respiration” OR “artificial ventilat*” OR “mechanical ventilat*” OR “NIV” OR “NPPV” OR “NIPPV” OR “positive pressure ventilat*”))
7 [NIV component]	5 OR 6
8	DE “Qualitative Methods”
9	TI ((("semi-structured" or semistructured or unstructured or informal or "in-depth" or indepth or "face-to-face" or structured or guide) n3 (interview* or discussion* or questionnaire*))) OR AB ((("semi-structured" or semistructured or unstructured or informal or "in-depth" or indepth or "face-to-face" or structured or guide) n3 (interview* or discussion* or questionnaire*)))
10	TI (focus group* or qualitative or ethnograph* or fieldwork or "field work" or "key informant") OR KW (focus group* or qualitative or ethnograph* or fieldwork or "field work" or "key informant")
11 [Qualitative component]	8 OR 9 OR 10
12 [Whole Search]	4 AND 7 AND 11



	Search ID#	Search Terms	Search Options	Actions
☐	S12	S4 AND S7 AND S11	**Expanders** - Apply equivalent subjects **Search modes** - Boolean/Phrase	View Results (8) | View Details | Edit
☐	S11	S8 OR S9 OR S10	**Expanders** - Apply equivalent subjects **Search modes** - Boolean/Phrase	View Results (156,023) | View Details | Edit
☐	S10	TI ((focus group* or qualitative or ethnograph* or fieldwork or "field work" or "key informant")) OR KW ((focus group* or qualitative or ethnograph* or fieldwork or "field work" or "key informant"))	**Expanders** - Apply equivalent subjects **Search modes** - Boolean/Phrase	View Results (54,454) | View Details | Edit
☐	S9	TI (((("semi-structured" or semistructured or unstructured or informal or "in-depth" or indepth or "face-to-face" or structured or guide) n3 (interview* or discussion* or questionnaire*)))) OR AB (((("semi-structured" or semistructured or unstructured or informal or "in-depth" or indepth or "face-to-face" or structured or guide) n3 (interview* or discussion* or questionnaire*))))	**Expanders** - Apply equivalent subjects **Search modes** - Boolean/Phrase	View Results (113,278) | View Details | Edit
☐	S8	DE "Qualitative Methods"	**Expanders** - Apply equivalent subjects **Search modes** - Boolean/Phrase	View Results (9,600) | View Details | Edit
☐	S7	S5 OR S6	**Expanders** - Apply equivalent subjects **Search modes** - Boolean/Phrase	View Results (1,575) | View Details | Edit
☐	S6	TI (((“non invasive ventilat*” OR “non-invasive ventilat*” OR “noninvasive ventilat*” OR “artificial respiration” OR “artificial ventilat*” OR “mechanical ventilat*” OR “NIV” OR “NPPV” OR “NIPPV” OR “positive pressure ventilat*”))) OR AB (((“non invasive ventilat*” OR “non-invasive ventilat*” OR “noninvasive ventilat*” OR “artificial respiration” OR “artificial ventilat*” OR “mechanical ventilat*” OR “NIV” OR “NPPV” OR “NIPPV” OR “positive pressure ventilat*”)))	**Expanders** - Apply equivalent subjects **Search modes** - Boolean/Phrase	View Results (1,433) | View Details | Edit
☐	S5	DE "Artificial Respiration"	**Expanders** - Apply equivalent subjects **Search modes** - Boolean/Phrase	View Results (261) | View Details | Edit
☐	S4	S1 OR S2 OR S3	**Expanders** - Apply equivalent subjects **Search modes** - Boolean/Phrase	View Results (19,335) | View Details | Edit
☐	S3	TI (((“motor neuron* disease” or “amyotrophic lateral sclerosis” or “MND” or “ALS”))) OR AB (((“motor neuron* disease” or “amyotrophic lateral sclerosis” or “MND” or “ALS”)))	**Expanders** - Apply equivalent subjects **Search modes** - Boolean/Phrase	View Results (13,413) | View Details | Edit
☐	S2	DE "Motor Neurons"	**Expanders** - Apply equivalent subjects **Search modes** - Boolean/Phrase	View Results (7,234) | View Details | Edit
☐	S1	DE "Amyotrophic Lateral Sclerosis"	**Expanders** - Apply equivalent subjects **Search modes** - Boolean/Phrase	View Results (4,419) | View Details | Edit

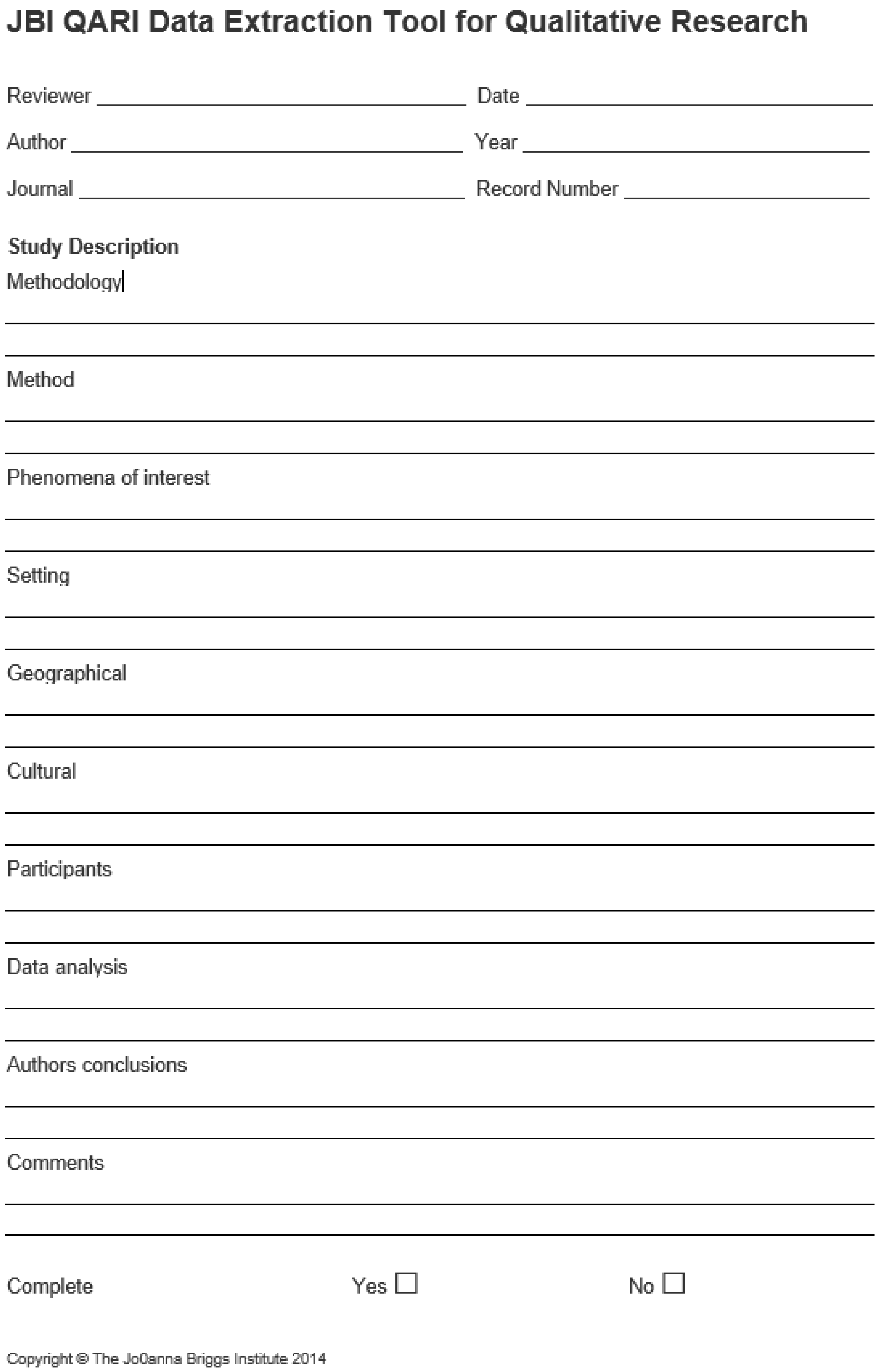
Search Results: 1 - 8 of 8

CINAHL	
1	(MH “Amyotrophic Lateral Sclerosis”)
2	(MH “Motor Neuron Diseases”)
3	TI ((“motor neuron* disease” or “amyotrophic lateral sclerosis” or “MND” or “ALS”)) OR AB ((“motor neuron* disease” or “amyotrophic lateral sclerosis” or “MND” or “ALS”))
4 [MND Component]	1 OR 2 OR 3
5	(MH “Respiration, Artificial”)
6	TI ((“non invasive ventilat*” OR “non-invasive ventilat*” OR “noninvasive ventilat*” OR “artificial respiration” OR “artificial ventilat*” OR “mechanical ventilat*” OR “NIV” OR “NPPV” OR “NIPPV” OR “positive pressure ventilat*”)) OR AB ((“non invasive ventilat*” OR “non-invasive ventilat*” OR “noninvasive ventilat*” OR “artificial respiration” OR “artificial ventilat*” OR “mechanical ventilat*” OR “NIV” OR “NPPV” OR “NIPPV” OR “positive pressure ventilat*”))
7 [NIV component]	5 OR 6
8	(MH “Qualitative Studies”)
9	TI ((("semi-structured" or semistructured or unstructured or informal or "in-depth" or indepth or "face-to-face" or structured or guide) n3 (interview* or discussion* or questionnaire*))) OR AB ((("semi-structured" or semistructured or unstructured or informal or "in-depth" or indepth or "face-to-face" or structured or guide) n3 (interview* or discussion* or questionnaire*)))
10	TI (focus group* or qualitative or ethnograph* or fieldwork or "field work" or "key informant") OR MW (focus group* or qualitative or ethnograph* or fieldwork or "field work" or "key informant")
11 [Qualitative component]	8 OR 9 OR 10
12 [whole search]	4 AND 7 AND 11



	Search ID#	Search Terms	Search Options	Actions
☐	S12	S4 AND S7 AND S11	**Expanders** - Apply equivalent subjects **Search modes** - Boolean/Phrase	View Results (20) View Details Edit
☐	S11	S8 OR S9 OR S10	**Expanders** - Apply equivalent subjects **Search modes** - Boolean/Phrase	View Results (216,746) View Details Edit
☐	S10	TI ((focus group* or qualitative or ethnograph* or fieldwork or "field work" or "key informant")) OR MW ((focus group* or qualitative or ethnograph* or fieldwork or "field work" or "key informant"))	**Expanders** - Apply equivalent subjects **Search modes** - Boolean/Phrase	View Results (165,870) View Details Edit
☐	S9	TI (((("semi-structured" or semistructured or unstructured or informal or "in-depth" or indepth or "face-to-face" or structured or guide) n3 (interview* or discussion* or questionnaire*)))) OR AB (((("semi-structured" or semistructured or unstructured or informal or "in-depth" or indepth or "face-to-face" or structured or guide) n3 (interview* or discussion* or questionnaire*))))	**Expanders** - Apply equivalent subjects **Search modes** - Boolean/Phrase	View Results (90,606) View Details Edit
☐	S8	(MH "Qualitative Studies")	**Expanders** - Apply equivalent subjects **Search modes** - Boolean/Phrase	View Results (120,957) View Details Edit
☐	S7	S5 OR S6	**Expanders** - Apply equivalent subjects **Search modes** - Boolean/Phrase	View Results (34,066) View Details Edit
☐	S6	TI (((“non invasive ventilat*” OR “non-invasive ventilat*” OR “noninvasive ventilat*” OR “artificial respiration” OR “artificial ventilat*” OR “mechanical ventilat*” OR “NIV” OR “NPPV” OR “NIPPV” OR “positive pressure ventilat*”))) OR AB (((“non invasive ventilat*��� OR “non-invasive ventilat*” OR “noninvasive ventilat*” OR “artificial respiration” OR “artificial ventilat*” OR “mechanical ventilat*” OR “NIV” OR “NPPV” OR “NIPPV” OR “positive pressure ventilat*”)))	**Expanders** - Apply equivalent subjects **Search modes** - Boolean/Phrase	View Results (20,946) View Details Edit
☐	S5	(MH "Respiration, Artificial")	**Expanders** - Apply equivalent subjects **Search modes** - Boolean/Phrase	View Results (22,614) View Details Edit
☐	S4	S1 OR S2 OR S3	**Expanders** - Apply equivalent subjects **Search modes** - Boolean/Phrase	View Results (10,735) View Details Edit
☐	S3	TI (((“motor neuron* disease” or “amyotrophic lateral sclerosis” or “MND” or “ALS”))) OR AB (((“motor neuron* disease” or “amyotrophic lateral sclerosis” or “MND” or “ALS”)))	**Expanders** - Apply equivalent subjects **Search modes** - Boolean/Phrase	View Results (9,243) View Details Edit
☐	S2	(MH "Motor Neuron Diseases")	**Expanders** - Apply equivalent subjects **Search modes** - Boolean/Phrase	View Results (1,365) View Details Edit
☐	S1	(MH "Amyotrophic Lateral Sclerosis")	**Expanders** - Apply equivalent subjects **Search modes** - Boolean/Phrase	View Results (4,211) View Details Edit

Appendix 1.3 JBI QARI Data Extraction Tool for Qualitative Research



JBI QARI Data Extraction Tool for Qualitative Research

Reviewer ______________________ Date ______________________

Author ______________________ Year ______________________

Journal ______________________ Record Number ______________________

Study Description

Methodology

____

____

Method

____

____

Phenomena of interest

____

____

Setting

____

____

Geographical

____

____

Cultural

____

____

Participants

____

____

Data analysis

____

____

Authors conclusions

____

____

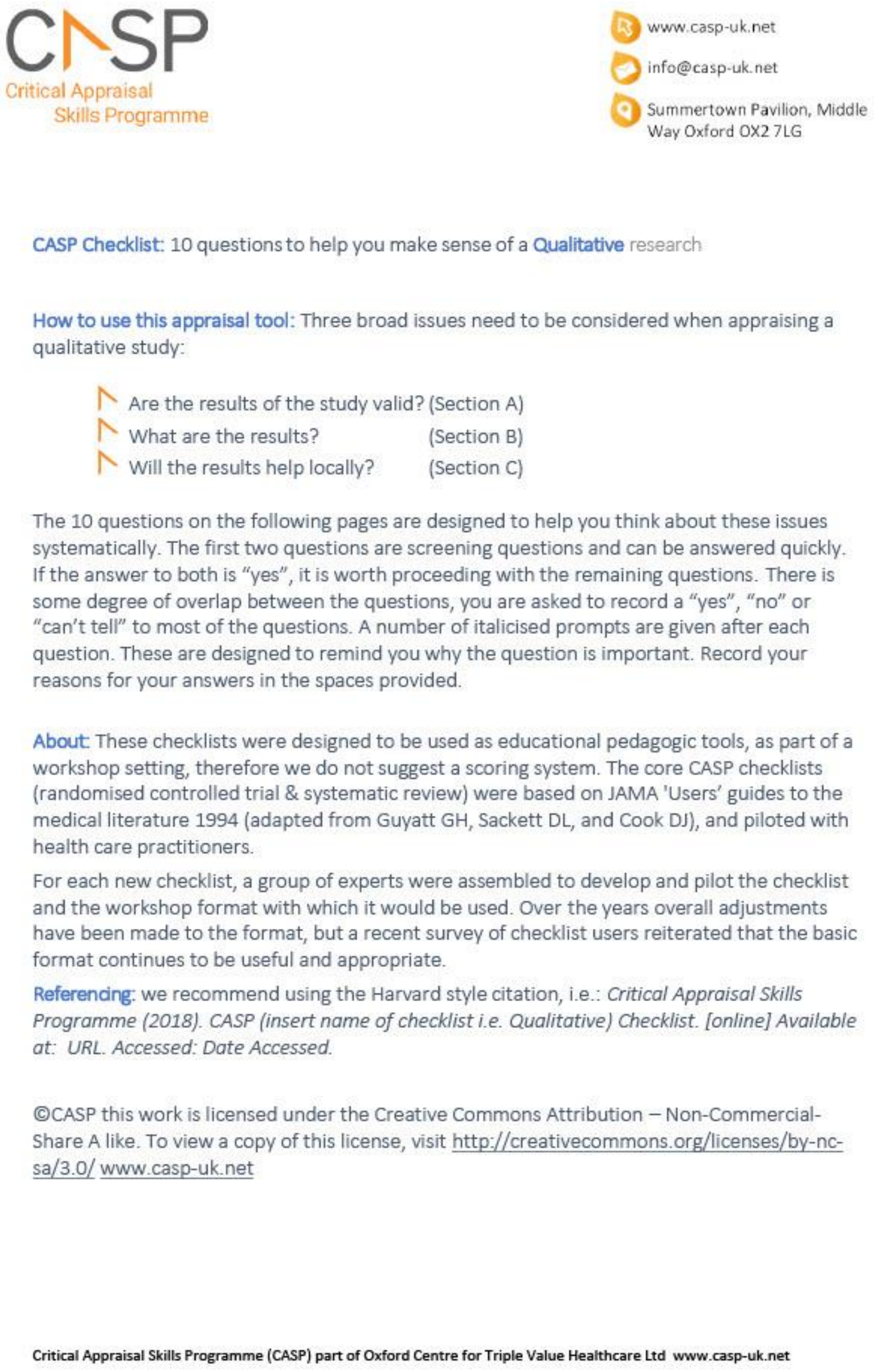
Comments

____

____

Complete Yes ☐ No ☐

Appendix 1.4 Critical Appraisal Skills Programme (CASP) Qualitative Checklist (2018)



CASP
Critical Appraisal
Skills Programme

www.casp-uk.net
info@casp-uk.net
Summertown Pavilion, Middle
Way Oxford OX2 7LG

CASP Checklist: 10 questions to help you make sense of a **Qualitative** research

How to use this appraisal tool: Three broad issues need to be considered when appraising a qualitative study:

- Are the results of the study valid? (Section A)
- What are the results? (Section B)
- Will the results help locally? (Section C)

The 10 questions on the following pages are designed to help you think about these issues systematically. The first two questions are screening questions and can be answered quickly. If the answer to both is "yes", it is worth proceeding with the remaining questions. There is some degree of overlap between the questions, you are asked to record a "yes", "no" or "can't tell" to most of the questions. A number of italicised prompts are given after each question. These are designed to remind you why the question is important. Record your reasons for your answers in the spaces provided.

About: These checklists were designed to be used as educational pedagogic tools, as part of a workshop setting, therefore we do not suggest a scoring system. The core CASP checklists (randomised controlled trial & systematic review) were based on JAMA 'Users' guides to the medical literature 1994 (adapted from Guyatt GH, Sackett DL, and Cook DJ), and piloted with health care practitioners.

For each new checklist, a group of experts were assembled to develop and pilot the checklist and the workshop format with which it would be used. Over the years overall adjustments have been made to the format, but a recent survey of checklist users reiterated that the basic format continues to be useful and appropriate.

Referencing: we recommend using the Harvard style citation, i.e.: *Critical Appraisal Skills Programme (2018). CASP (insert name of checklist i.e. Qualitative) Checklist. [online] Available at: URL. Accessed: Date Accessed.*

Critical Appraisal Skills Programme (CASP) part of Oxford Centre for Triple Value Healthcare Ltd www.casp-uk.net

Paper for appraisal and reference: ....

Section A: Are the results valid?

1. Was there a clear statement of the aims of the research?

Yes ☐
Can't Tell ☐
No ☐

HINT: Consider
- what was the goal of the research
- why it was thought important
- its relevance

Comments:

2. Is a qualitative methodology appropriate?

Yes ☐
Can't Tell ☐
No ☐

HINT: Consider
- If the research seeks to interpret or illuminate the actions and/or subjective experiences of research participants
- Is qualitative research the right methodology for addressing the research goal

Comments:

Is it worth continuing?

3. Was the research design appropriate to address the aims of the research?

Yes ☐
Can't Tell ☐
No ☐

HINT: Consider
- if the researcher has justified the research design (e.g. have they discussed how they decided which method to use)

Comments:

4. Was the recruitment strategy appropriate to the aims of the research?

Yes ☐
Can't Tell ☐
No ☐

HINT: Consider

- If the researcher has explained how the participants were selected
- If they explained why the participants they selected were the most appropriate to provide access to the type of knowledge sought by the study
- If there are any discussions around recruitment (e.g. why some people chose not to take part)

Comments:

5. Was the data collected in a way that addressed the research issue?

Yes ☐
Can't Tell ☐
No ☐

HINT: Consider

- If the setting for the data collection was justified
- If it is clear how data were collected (e.g. focus group, semi-structured interview etc.)
- If the researcher has justified the methods chosen
- If the researcher has made the methods explicit (e.g. for interview method, is there an indication of how interviews are conducted, or did they use a topic guide)
- If methods were modified during the study. If so, has the researcher explained how and why
- If the form of data is clear (e.g. tape recordings, video material, notes etc.)
- If the researcher has discussed saturation of data

Comments:

6. Has the relationship between researcher and participants been adequately considered?

Yes ☐
Can't Tell ☐
No ☐

HINT: Consider

- If the researcher critically examined their own role, potential bias and influence during (a) formulation of the research questions (b) data collection, including sample recruitment and choice of location
- How the researcher responded to events during the study and whether they considered the implications of any changes in the research design

Comments:

Section B: What are the results?

7. Have ethical issues been taken into consideration?

Yes ☐
Can't Tell ☐
No ☐

HINT: Consider

- If there are sufficient details of how the research was explained to participants for the reader to assess whether ethical standards were maintained
- If the researcher has discussed issues raised by the study (e.g. issues around informed consent or confidentiality or how they have handled the effects of the study on the participants during and after the study)
- If approval has been sought from the ethics committee

Comments:

8. Was the data analysis sufficiently rigorous?

Yes ☐
Can't Tell ☐
No ☐

HINT: Consider

- If there is an in-depth description of the analysis process
- If thematic analysis is used. If so, is it clear how the categories/themes were derived from the data
- Whether the researcher explains how the data presented were selected from the original sample to demonstrate the analysis process
- If sufficient data are presented to support the findings
- To what extent contradictory data are taken into account
- Whether the researcher critically examined their own role, potential bias and influence during analysis and selection of data for presentation

Comments:

9. Is there a clear statement of findings?

Yes ☐
Can't Tell ☐
No ☐

HINT: Consider whether

- If the findings are explicit
- If there is adequate discussion of the evidence both for and against the researcher's arguments
- If the researcher has discussed the credibility of their findings (e.g. triangulation, respondent validation, more than one analyst)
- If the findings are discussed in relation to the original research question

Comments:

Section C: Will the results help locally?

10. How valuable is the research?

HINT: Consider
- If the researcher discusses the contribution the study makes to existing knowledge or understanding (e.g. do they consider the findings in relation to current practice or policy, or relevant research-based literature
- If they identify new areas where research is necessary
- If the researchers have discussed whether or how the findings can be transferred to other populations or considered other ways the research may be used

Comments:

Appendix 1.5 Seven Steps to Meta-Ethnography (Noblit & Hare, 1988)

1. Getting Started	•Identify an area of interest within qualitative research
2. Deciding what is relevant to the initial interest	•Define the focus of the synthesis •Identify eligible studies •Appraise quality of studies
3. Reading the studies	•Become familiar with the themes in each study
4. Determining how the studies are related	•Generate a list of themes for each study •Examine relationships between concepts/themes
5. Translating studies into one another	•Use these themes to translate the studies into one another
6. Synthesising translations	•Studies are viewed as a whole •Reciprocal and refutational synthesis, line of argument synthesis
7. Expressing the synthesis	•Write up the synthesis following eMERGE and/or PRISMA reporting guidlines

Appendix 1.6 Quality Appraisal Responses for all Included Studies

Study	**1**	**2**	**3**	**4**	**5**	**6**	**7**	**8**	**9**	**10**	**Comments**
Ando et al. (2014) “Experience of…”	Y	Y	Y	Y	Y	Y	Y	Y	Y	Y	Same study
Ando et al. (2014) “Why don’t they…”	Y	Y	Y	Y	Y	Y	Y	Y	Y	Y	
Baxter et al. (2013) “The impact on…”	Y	Y	Y	Y	Y	N	Y	Y	Y	Y	Same study
Baxter et al. (2013) “The initiation of…”	Y	Y	Y	Y	Y	N	Y	Y	Y	Y	
Baxter et al. (2013) “The use of…”	Y	Y	Y	Y	Y	N	CT	Y	Y	Y	
Greenaway et al. (2015)	Y	Y	Y	Y	Y	N	Y	Y	Y	Y	
Lemoignan & Ells (2010)	Y	Y	Y	Y	Y	Y	Y	Y	Y	Y	
Sundling et al. (2009)	Y	Y	Y	Y	Y	Y	Y	Y	Y	CT	
Winther et al. (2020)	Y	Y	Y	Y	Y	Y	Y	CT	Y	Y	
Young et al. (1994)	Y	Y	Y	Y	Y	Y	CT	N	CT	Y	

Response: Yes (Y), No (N), Can’t tell (CT).

See Appendix 1.3 for CASP questions

Appendix 1.7 Themes identified from reciprocal translation during meta-ethnography.

Theme	Reciprocal Translation	Studies
1.1 Desire to live and acceptance	6 out of 10	Ando et al. (2014a) Baxter et al. (2013b) Greenaway et al. (2015) Lemoignan & Ells (2010) Winther et al. (2020) Young et al. (1994)
1.2 Reluctance and resistance	5 out of 10	Ando et al. (2014a) Ando et al. (2014b) Greenaway et al. (2015) Lemoignan & Ells (2010) Young et al. (1994)
2.1 Initial discomfort	6 out of 10	Ando et al. (2014a) Ando et al. (2014b) Baxter et al. (2013a) Baxter et al. (2013b) Sundling et al. (2009) Young et al. (1994)
1.2 Perceived benefits	7 out of 10	Ando et al. (2014a) Baxter et al. (2013a) Baxter et al. (2013b) Baxter et al. (2013c) Lemoignan & Ells (2010) Sundling et al. (2009) Young et al. (1994)
3. Impact of continuing deterioration	5 out of 10	Baxter er al. (2013a) Baxter et al. (2013c) Lemoignan & Ells (2010) Greenaway et al. (2015) Winther et al. (2020)
4.1 Fear of dependence and loss of autonomy	8 out of 10	Ando et al. (2014a) Ando et al. (2014b) Baxter et al. (2013a) Baxter et al. (2013c) Lemoignan & Ells (2010) Sundling et al. (2009) Winther et al. (2020) Young et al. (1994)
4.2 Fears influencing decision making	5 out of 10	Ando et al. (2014b) Baxter et al. (2013b) Lemoignan & Ells (2010) Greenaway et al. (2015) Sundling et al. (2009)
5. Experience of healthcare professionals	8 out of 10	Ando et al. (2014b) Baxter et al. (2013)a Baxter et al. (2013b) Lemoignan & Ells (2010) Greenaway et al. (2015)

		Sundling et al. (2009) Winther et al. (2020)

Appendix 2.1 Adapted Motor Neurone Disease Distress Thermometer and Concerns Checklist

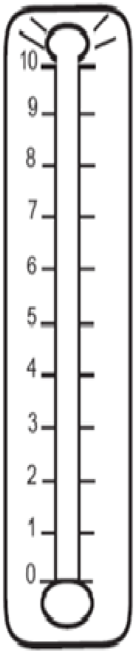
NHS Ayrshire & Arran

Motor Neurone Disease Concerns Checklist

Patient Name: …………………………

Date completed ……………………….

1. Circle the number from 0 (zero) to 10 (ten) that best describes how much distress you have felt in the past week, including today.



2. Please tick any of the following that has been a cause of distress for you in the past week, including today.

Physical problems
☐ Appearance
☐ Bathing/dressing
☐ Breathing
☐ Bladder/bowel
☐ Choking
☐ Eating/drinking
☐ Fatigue/tiredness
☐ Movement/getting around
☐ Pain
☐ Sexual
☐ Skin
☐ Sleeping
☐ Speech
☐ Salvia management

Practical problems
☐ Child care
☐ Finances
☐ Housing
☐ Transport/Driving
☐ Work/activities
☐ Legal issues
☐ Aids and adaptations

Cognitive problems
☐ Concentration
☐ Communication
☐ Memory
☐ Making decisions

Relationships
☐ Dealing with family or friends
☐ Dealing with staff/carers
☐ Roles and responsibilities

Emotional problems
☐ Worry/fear/anxiety
☐ Sadness/depression
☐ Sudden crying/laughing
☐ Anger
☐ Guilt
☐ Denial

Behavioural problems
☐ Changes in behaviour/personality
☐ Loss of interest or motivation
☐ Loss of interest in how other feel
☐ Change in food preferences

Spiritual
☐ Spiritual concerns
☐ Religious concerns

Questions about illness or treatment:....................................
....
....
....

3. If there are several concerns please identify which three would help you most to discuss at this time?

Concern 1.	Concern 2.	Concern 3.

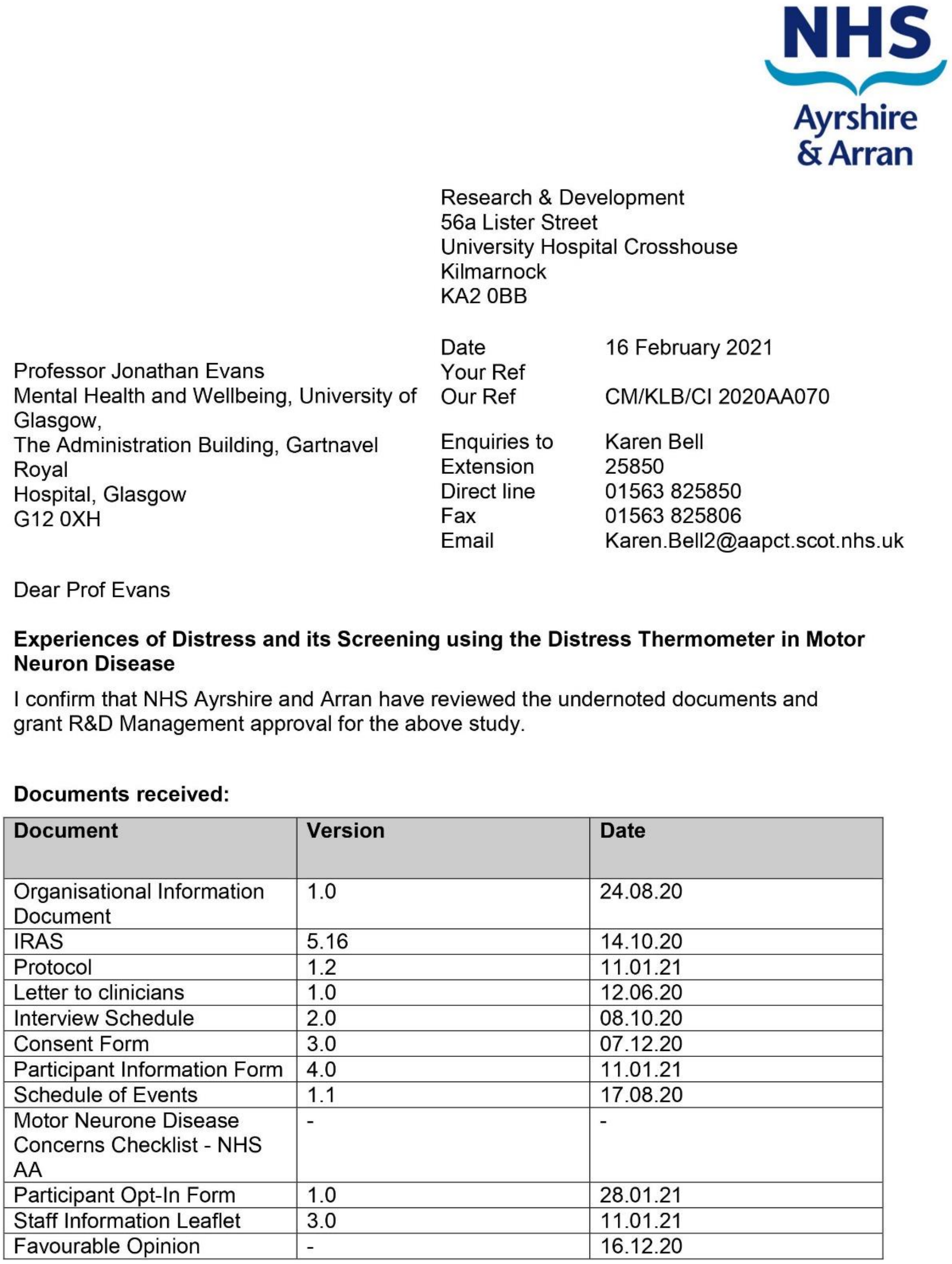
MND Concerns Checklist – Management

	Briefly describe the Patient's concern.	Any immediate action taken e.g. information clarified, verbal or written information given, referred to see other health care professional	Any next steps required? e.g. Referral to other service, speak to MDT, seek advice	Who is responsible for the next step or follow up	Date of review follow up
Concern 1					
Concern 2					
Concern 3					

Any further comments

Print Name: ______________________ Signature: ________________ Contact Number: ________________Date: ___________

Appendix 2.2 NHS Ayrshire and Arran Research and Development Approval



Research & Development
56a Lister Street
University Hospital Crosshouse
Kilmarnock
KA2 0BB

Date	16 February 2021
Your Ref	
Our Ref	CM/KLB/CI 2020AA070
Enquiries to	Karen Bell
Extension	25850
Direct line	01563 825850
Fax	01563 825806
Email	Karen.Bell2@aapct.scot.nhs.uk

Professor Jonathan Evans
Mental Health and Wellbeing, University of Glasgow,
The Administration Building, Gartnavel Royal
Hospital, Glasgow
G12 0XH

Dear Prof Evans

Experiences of Distress and its Screening using the Distress Thermometer in Motor Neuron Disease

I confirm that NHS Ayrshire and Arran have reviewed the undernoted documents and grant R&D Management approval for the above study.

Documents received:

Document	Version	Date
Organisational Information Document	1.0	24.08.20
IRAS	5.16	14.10.20
Protocol	1.2	11.01.21
Letter to clinicians	1.0	12.06.20
Interview Schedule	2.0	08.10.20
Consent Form	3.0	07.12.20
Participant Information Form	4.0	11.01.21
Schedule of Events	1.1	17.08.20
Motor Neurone Disease Concerns Checklist - NHS AA	-	-
Participant Opt-In Form	1.0	28.01.21
Staff Information Leaflet	3.0	11.01.21
Favourable Opinion	-	16.12.20

The terms of approval state that the investigator authorised to undertake this study within NHS Ayrshire & Arran is: -

- Rachel King, NHS Ayrshire & Arran

With additional investigators:-

- Dr Luke Williams, NHS Ayrshire & Arran

The sponsors for this study are University of Glasgow.

This approval letter is valid until 30 January 2022.

Regular reports of the study require to be submitted. Your first report should be submitted to Dr K Bell, Research & Development Manager in 12 months time and subsequently at yearly intervals until the work is completed.

Please note that as a requirement of this type of study your name, designation, work address, work telephone number, work e-mail address, work related qualifications and whole time equivalent will be held on the Scottish National Research Database so that NHS R&D staff in Scotland can access this information for purposes related to project management and report monitoring.

In addition approval is granted subject to the following conditions: -

- All research activity must comply with the standards detailed in the UK Policy Framework for Health and Social Care Research http://beta.hra.nhs.uk/planning-and-improving-research/policies-standards-legislation/uk-policy-framework-health-social-care-research and appropriate statutory legislation. It is your responsibility to ensure that you are familiar with these, however please do not hesitate to seek further advice if you are unsure.

- Recruitment figures must be submitted to R&D on a monthly basis. If recruitment figures are not received timeously you will be contacted by a member of the R&D team to provide this data.

- You are required to comply with Good Clinical Practice (ICH-GCP guidelines may be found at www.ich.org/LOB/media/MEDIA482.pdf), Ethics Guidelines, Health & Safety Act 1999, General Data Protection Regulation (GDPR) and Data Protection Act 2018.

- If any amendments are to be made to the study protocol and or the Research Team the Researcher must seek Ethical and Management Approval for the changes before they can be implemented.

- The Researcher and NHS Ayrshire and Arran must permit and assist with any monitoring, auditing or inspection of the project by the relevant authorities.

- The NHS Ayrshire and Arran Complaints Department should be informed if any complaints arise regarding the project and the R&D Department must be copied into this correspondence.

- The outcome and lessons learnt from complaints must be communicated to funders, sponsors and other partners associated with the project.

- As custodian of the information collated during this research project you are responsible at all times for ensuring the security of all personal information collated in line with NHS Scotland policies on information assurance and security, until the secure destruction of these data. The retention time periods for such data should comply with the requirements of the Scottish Government Records Management: NHS Code Of Practice. Under no circumstances should personal data be stored on any unencrypted removable media e.g. laptop, USB or mobile device (for further information and guidance please contact the Information Governance Team based at University Hospital Crosshouse 01563 825831 or 826813).

If I can be of any further assistance please do not hesitate to contact me. On behalf of the department, I wish you every success with the project.

Yours sincerely

[redacted]

Dr Crawford McGuffie
Medical Director

c.c. Dr Colette Montgomery Sardar, University of Glasgow (sponsor contact)
Lesley Douglas, Finance, Ailsa Hospital
Information Governance, NHS Ayrshire & Arran
Rachel King, NHS Ayrshire & Arran
Dr Luke Williams, NHS Ayrshire & Arran

Appendix 2.3 Ethical Approval from West of Scotland REC 3

WoSRES
West of Scotland Research Ethics Service

NHS
Greater Glasgow
and Clyde

Ms Rachel King
Mental Health and Wellbeing, University of Glasgow,
The Administration Building, Gartnavel Royal
Hospital, Glasgow
G12 0XH

West of Scotland REC 3
Research Ethics
Clinical Research and Development
Dykebar Hospital
Grahamston Road
Paisley PA2 7DE

Date	10 March 2021
Direct line	0141 314 0211
E-mail	WoSREC3@ggc.scot.nhs.uk

Dear Ms King

Study title:	**Experiences of Distress and its Screening using the Distress Thermometer in Motor Neuron Disease**
REC reference:	**20/WS/0157**
Amendment number:	**281506Amend1.2Substantial 02.03.2021 (REC Ref AM02)**
Amendment date:	**02 March 2021**
IRAS project ID:	**281506**

The above amendment was reviewed by the Sub-Committee in correspondence.

Ethical opinion

The members of the Committee taking part in the review gave a favourable ethical opinion of the amendment on the basis described in the notice of amendment form and supporting documentation.

Approved documents

The documents reviewed and approved at the meeting were:

Document	*Version*	*Date*
Completed Amendment Tool [IRAS281506 Amendment Tool V1.2Substantial 02032021]	281506Amend1.2Substantial 02.03.2021 (REC Ref AM02)	02 March 2021
Covering letter on headed paper [Cover Letter Re Amendment Tool V1.2Substantial 02032021]		02 March 2021
GP/consultant information sheets or letters [Staff Information Leaflet V3.1 02032021]	3.1	02 March 2021
Other [Participant Opt-In Form V1 28012021]	1	28 January 2021

Membership of the Committee

The members of the Committee who took part in the review are listed on the attached sheet.

Working with NHS Care Organisations

Sponsors should ensure that they notify the R&D office for the relevant NHS care organisation of this amendment in line with the terms detailed in the categorisation email issued by the lead nation for the study.

Amendments related to COVID-19

We will update your research summary for the above study on the research summaries section of our website. During this public health emergency, it is vital that everyone can promptly identify all relevant research related to COVID-19 that is taking place globally. If you have not already done so, please register your study on a public registry as soon as possible and provide the HRA with the registration detail, which will be posted alongside other information relating to your project.

Statement of compliance

The Committee is constituted in accordance with the Governance Arrangements for Research Ethics Committees and complies fully with the Standard Operating Procedures for Research Ethics Committees in the UK.

HRA Learning

We are pleased to welcome researchers and research staff to our HRA Learning Events and online learning opportunities– see details at: https://www.hra.nhs.uk/planning-and-improving-research/learning/

IRAS Project ID - 281506: **Please quote this number on all correspondence**

Yours sincerely

[redacted]

for
Dr Anne-Louise Cunnington
Chair

Enclosures:	*List of names and professions of members who took part in the review*
Copy to:	*Ms Rachel King*

West of Scotland REC 3

Attendance at Sub-Committee of the REC meeting on 10 March 2021

Committee Members:

Name	*Profession*	*Present*	*Notes*
Mr Daniel Boyle	Scriptwriter	Yes	
Dr Kenneth Harden	Retired GP	Yes	
Mr Ben Parkinson	Lecturer in Nursing and Vice Chair (Sub-Committee Chair)	Yes	

Appendix 2.4 Participant Consent Form

University of Glasgow
Institute of Health & Wellbeing

NHS
Ayrshire & Arran

Consent Form

Title of project: Experiences of Distress and its Screening using the Distress Thermometer and Concerns Checklist in Motor Neurone Disease

Name of researcher: Rachel King

Patient identification number for this study:

Please initial the box

1. I confirm that I have read and understood the Participant Information sheet version No: (date) for the above study. ☐
2. I have had the opportunity to think about the information and ask questions, and understand the answers I have been given. ☐
3. I understand that my participation in the above study is voluntary and confidential and that I am free to leave the study at any time without giving any reason and without my current or future treatment being affected. ☐
4. I understand that if I withdraw from the study, my data collected up to that point will be retained and used for the remainder of the study but no new research procedures will be carried out. ☐
5. I give permission for the interview to be audio recorded. ☐
6. I agree that you may inform the Motor Neuron Disease Clinic that I have agreed to participate in the study. ☐
7. I understand that if any issues arise during the interview, the researcher will inform me of this and discuss the possibility of sharing this with the Motor Neurone Disease team. ☐

Version 3 07/12/2020

8. I understand that any information that I provide may be quoted during dissemination of the study research and that this will be done anonymously and with all identifiable information removed. ☐

9. I agree that my name, contact details and data described in the information sheet will be kept for the purposes of this research project. ☐

10. I confirm that I agree to the way my data will be collected and processed and that data will be stored for up to 10 years in University archiving facilities in accordance with relevant Data Protection policies and regulations. ☐

11. I understand that all data and information I provide will be kept confidential and will be seen only by study researchers and regulators whose job it is to check the work of researchers. ☐

12. Optional: I would like to receive a copy of the findings from the study. Please only tick this box if you would like a copy of the findings. This is not a requirement of participating in the study. ☐

13. I agree to take part in the study. ☐

Participant Name	**Date**	**Signature**
…………………………………………	……/……/……….	…………………………………………
Researcher	**Date**	**Signature**
…………………………………………	……/……/……….	…………………………………………

Thank you for agreeing to take part in this research

Version 3 07/12/2020

Appendix 2.5 Staff Information Leaflet

Version 3.1 02/03/2021

University of Glasgow
Institute of Health & Wellbeing

NHS Ayrshire & Arran

Staff Information Leaflet

Experiences of Distress and its Screening using the Distress Thermometer in Motor Neurone Disease

My name is Rachel King and I am a Trainee Clinical Psychologist with the University of Glasgow. As part of my Doctorate in Clinical Psychology I am undertaking a research project investigating the experience of distress for people who live with Motor Neurone Disease (MND), and the extent to which this relates to their experiences of either using or viewing the Distress Thermometer and Concerns Checklist. These tools are used to guide health conversations within the Motor Neurone Disease Clinic when the clinic is operating normally. As this is not the case currently, not all potential participants will have had the chance to go through these with a clinician during their appointments.

What is the purpose of the study?

The purpose of the study is to increase our understanding of the experience of distress in people who are living with a diagnosis of MND. It is hoped that the study will provide us with a better understanding of the experiences and difficulties that people with MND face. It is also hoped that it will help to inform us about how well the Distress Thermometer and Concerns Checklist capture this experience for patients, and whether this can be improved.

What does the study involve?

The study involves interviewing participants who have a diagnosis of Motor Neurone Disease and attend the MND Clinic in NHS Ayrshire and Arran. Participants must meet the following criteria:

- Diagnosis of Motor Neurone Disease
- Registered and seen within the Motor Neurone Disease Clinic
- Cognitive capacity to communicate informed consent
- Aged 18 years or over

Unfortunately, it is beyond the scope of the current study to include people who have communication difficulties which would be considered too difficult to understand via audio recording. It is hoped that this could be the focus of further research as communication difficulties would be expected to increase distress that people experience, and this is a group who are often excluded from research studies.

Patients who meet the criteria above and who would like to participate will take part in a semi-structured interview. Interviews will be expected to last between 1-1.5 hours, but timings and numbers of interviews conducted will be flexible. Participants will be given the option of multiple shorter interviews and longer interviews will be conducted if this is required. In all cases, breaks can be taken by participants as and when needed. Interviews will be conducted using video conferencing. Patients will be asked to give consent verbally before they participate, and they can opt-out of participation at any time. No patient identifiable information will be stored.

How can I help?

It is hoped that you could identify patients from your caseload who meet the above inclusion criteria, based on your clinical judgement. I would then ask that you make your patient aware of the study during your next contact. At this point, you would provide verbal information and provide patients with an information sheet about the study, or send this out to patients if you are unable to meet with them in person. You could also share the information sheet over Attend Anywhere if you are using this to meet with patients. If your patient agrees to participate at this stage, please complete the Opt-In Form with the patient's contact details and pass this onto me. I will then contact the participant by telephone to answer any study related questions that they may have and to arrange and appointment for an interview.

What happens next?

I will contact your patient and explain the details of the study and answer any questions that they have about the study. I will remind your patient that their participation in the study is optional and that they can withdraw from the study at any time. If they agree to participate at this stage, I will arrange an appointment with them to conduct the interview. I will then contact you to let you know that your patient has consented to participate, and the date of the interview.

Do people have to take part?

No, it is completely up to individuals whether they would like to take part or not. Even if people do decide to take part, they can decide to withdraw from the study at a time if they change their mind about participating.

What happens to the information?

The audio recordings will be stored on an encrypted laptop and will be deleted as soon as they have been transcribed. All transcriptions will be anonymised, and pseudonyms will be used. Transcriptions will be stored within the secure Glasgow University network once the study has been completed. Data will be stored here for ten years in accordance with the Data Protection Act and will be destroyed after this period.

Any personal information (such as your patient's name and phone number) will be known to researcher but will be saved separately from audio recordings and transcriptions. This information will be stored in a locked filing cabinet within the MND clinic and will be destroyed once the study is complete.

The results will be written up as part of a Clinical Psychology doctoral thesis and will be published on the Glasgow University thesis website. The research will be shared with interested participants and members of MND clinic staff and may be submitted for publication. Some direct quotes from patient interviews may be included in reports and publications, but all information will be anonymised so that it is not possible for anyone to be personally identified from this information.

What are the possible benefits of taking part?

NICE guidelines (2016) state that people with MND should be given the opportunity to take part in research. Participants with MND may be interested in sharing their experiences to help improve the understanding of distress in MND how this relates to how psychological issues are screened for in clinics. However, participants themselves may receive no direct benefit from participating in this research.

Are there any disadvantages to patients taking part?

Due to the nature of the topic, it is possible that participants may become upset or distressed during or following interviews. During interviews, the researcher will frequently check in with participants to check for fatigue or distress and ensure they are happy to continue. The researcher will allow time at the end of interviews for participants to reflect on how they are feeling. All participants will have the opportunity to contact a nurse or psychologist from the MNDC for support after their interview and relevant clinicians will be aware of when interviews are taking place.

Who has reviewed this study?

This study has been reviewed by NHS Ayrshire and Arran Research & Development Department and has been reviewed by Research Ethics Committee 3, West of Scotland to ensure that the study is being conducted correctly.

What if something goes wrong?

If you have any concerns about the study or the way it is conducted, please contact Dr Luke Williams, Dr Luke Williams, 1st Floor, Ayrshire Central Hospital, Kilwinning Road, KA12 8SS. Email: Luke.Williams@aapct.scot.nhs.uk. Tel: 01294 322057

Any questions?

Please do not hesitate to contact me should you have any questions about the study. Or if you would like any further information. Below is contact information for all researchers involved in the study:

Researcher(s) Contact Details:
Rachel King, Trainee Clinical Psychologist Institute of Mental Health & Wellbeing, University of Glasgow Administration Building, 1st Floor Gartnavel Royal Hospital 1055 Great Western Road Glasgow G12 0XH

Email: [redacted]
Professor Jonathan Evans Institute of Mental Health & Wellbeing, University of Glasgow Administration Building, 1st Floor Gartnavel Royal Hospital 1055 Great Western Road Glasgow G12 0XH Email: Jonathan.Evans@glasgow.ac.uk Tel: 0141 211 3978
Dr Luke Williams Clinical Psychologist 1st Floor Ayrshire Central Hospital Kilwinning Road KA12 8SS Email: Luke.Williams@aapct.scot.nhs.uk Tel: 01294 322057

Appendix 2.6 Participant Opt-In Form

Version 1 28.01.2021

University of Glasgow

Institute of Health & Wellbeing

NHS Ayrshire & Arran

Participant Opt-In Form

I agree to being contacted by researchers regarding the study entitled **Experiences of Distress and its Screening using the Distress Thermometer and Concerns Checklist in Motor Neurone Disease.** My clinician at the MND clinic has provided me with information about the study. I understand that they will be in contact to discuss the study with me but I am under no obligation to take part. I have agreed with my clinician to pass on the following contact details. I understand that if I would like to participate in the study, information will be sent out to me via post to my address.

Name: ____

Telephone number: ____

Address (including postcode): ____

____

Please return to Rachel King by NHS internal mail:

Rachel King, Trainee Clinical Psychologist, Medical Paediatrics, Crosshouse Hospital, Kilmarnock, KA2 0BE

Appendix 2.7 Participant Information Leaflet

Version 4 11/01/2021

University of Glasgow

Institute of Health & Wellbeing

NHS Ayrshire & Arran

Participant Information Leaflet

Experiences of Distress and its Screening using the Distress Thermometer and Concerns Checklist in Motor Neurone Disease

We would like to invite you to take part in a research study. To help you to decide whether or not you would like to take part, it is important that you understand what participating would involve for you. Please take your time and read the following information carefully.

Who is conducting the research?

The research is being carried out by Rachel King, who is a Trainee Clinical Psychologist at the University of Glasgow. The research is being supervised by Professor Jonathan Evans, Professor of Applied Neuropsychology from the University of Glasgow, and Dr Luke Williams, Clinical Psychologist from the NHS Ayrshire and Arran Neuropsychology Service (including the MND Clinic).

What is the purpose of the study?

The purpose of the study is to increase our understanding of the experience of distress in people who are living with a diagnosis of MND. The study is being carried out as part of the requirements of the Doctorate in Clinical Psychology training course at the University of Glasgow.

The study will involve talking to people who have a diagnosis of MND about their experiences of living with the illness. The study will also explore how people experience tools which are used to screen for distress in the MND Clinic in Ayrshire, and the extent to which this matches up with the experiences of individuals. These tools are the Distress Thermometer and the Concerns Checklist, and you may or may not have come across these at your clinic appointments. Part of the study will involve going over these tools with the researcher so that everyone has the chance to discuss whether or not they feel that these tools reflect their experience.

It is hoped that the study will provide us with a better understanding of the experiences and difficulties that people with MND face. It is also hoped that it will help to inform us about how well the Distress Thermometer and Concerns Checklist capture these experiences and whether these tools can be improved.

Why have I been invited?

We are inviting people who have a diagnosis of Motor Neurone Disease and who are currently attending the Motor Neurone Disease Clinic within Ayrshire and Arran. We believe that you meet these criteria and that is why we have invited you to take part.

What does taking part involve?

If you decide to take part in the study, you will be asked to:

1. Let the clinician you are seeing at the MND clinic know that you are interested in learning more about the study, and are happy to be contacted by Rachel King, who will then telephone you.
2. Rachel can provide you with more information about the study and answer any questions that you may have. If you decide that you would like to take part, she will arrange an appointment time with you. The appointments will take place over video conferencing software.
3. Before you begin the interview, Rachel will ask you to provide consent to say that you agree to take part in the study. This will be sent out to you prior to interview. If it is difficult for you to sign a consent form, you can have someone sign for you on your behalf or give verbal consent before we begin the interview. If you provide verbal consent, this will be recorded.
4. Your interview will last between an hour and an hour and a half and will be an informal discussion. Rachel will ask you some questions about your illness and how it has impacted on your life.

You are welcome to have someone you know present at the interview. You can take a break at any time during the interview and you don’t have to answer any questions that you don’t want to. Although the interview is expected to last between an hour and an hour and a half, these timings can be flexible. For example, if you would rather complete the interview in multiple, shorter sessions then we could arrange this.

The interviews will be audio recorded to make sure that what is written down in the study exactly matches what people participating in the study say. The research paper may include some quotes from your interview, but these will be anonymised to make sure that you cannot be identified from these. For example, if you mention the names of people or places then this information will be removed, and pseudonyms will be used.

Do I have to take part?

No. Your decision to take part or not is completely up to you. If you would like to take part, you will be asked to sign a consent form or give verbal consent before your interview, so that there is a record of your consent.

You can leave the study at any point without giving a reason. If you do choose to leave the study after you have begun to participate information you have provided up to the point that you withdrew will be retained but no new research procedures will be carried out. If you decide to leave the study, this will not affect the care or support that you receive now, or in the future.

What happens to the information?

We will inform clinicians whom you see at the MND clinic so that they are aware that you have chosen to take part in the study. If any issues arise during the interview, I will discuss this with you and make you aware of the possibility of sharing this information with the MND team.

The audio recordings will be stored on an encrypted laptop and will be deleted from the audio recorder as soon as they have been transferred to the encrypted laptop. Audio recordings will be deleted from the laptop at the end of the study. . All transcriptions will be anonymised, and pseudonyms will be used. Transcriptions will be stored within the secure Glasgow University network once the study has been completed. Data will be stored here for ten years in accordance with the Data Protection Act 2018 and the General Data Protection Regulation 2018, and will be destroyed after this period. The data that we collect from you will only be used for this study and will not be re-used in any other research.

Any personal information (such as your name and phone number) will be known to researcher but will be saved separately from audio recordings and transcriptions. This information will be stored in a locked filing cabinet within the MND clinic and will be destroyed once the study is complete.

The University of Glasgow is the sponsor for this study based in the United Kingdom. We will be using information from you in order to undertake this study and will act as the data controller for this study. This means that we are responsible for looking after your information and using it properly. The University of Glasgow will keep identifiable information about you for 10 years after the study has finished.

Your rights to access, change or move your information are limited, as we need to manage your information in specific ways in order for the research to be reliable and accurate. If you withdraw from the study, we will keep the information about you that we have already obtained. To safeguard your rights, we will use the minimum personally identifiable information possible.

You can find out more about how we use your information at: https://ico.org.uk/for-organisations/guide-to-data-protection/guide-to-the-general-data-protection-regulation-gdpr/

Your identifiable information might be shared with people who check on behalf of the study sponsor that the study is done properly.

The results of this study will be written up as part of a Clinical Psychology doctoral thesis and will be published on the Glasgow University thesis website. The research may also be submitted for publication in medical journals. Some direct quotes from your interviews may be included in reports and publications, but all information will be anonymised so that it is not possible for anyone to be personally identified from this information. If you would like to receive a copy of the findings from the study then please let Rachel know. You will then be sent a copy when the report is complete.

What are the possible benefits of taking part?

Taking part may not be of direct benefit to you. You may be interested in sharing your experiences to help improve the understanding of distress in MND how this relates to how psychological issues are screened for in clinics. It is hoped that this study may help to improve this experience for people with MND.

What are the potential risks and disadvantages of taking part?

There are no foreseen risks to taking part in the current study. We understand that due to the nature of the topic, it may be difficult or upsetting to answer some questions. It may also be quite tiring, and you will be able to do the interview in smaller chunks if this would feel easier for you. During interviews, the researcher will frequently check in with you to check that you are they are happy to continue. We will allow time at the end of interviews for you to reflect on how they are feeling. You will have the opportunity to contact a nurse or psychologist from the MNDC for support after their interview and relevant clinicians will be aware of when interviews are taking place.

Who has reviewed this study?

This study has been reviewed by NHS Ayrshire and Arran Research & Development Department and has been reviewed by an NHS Ethics Committee to ensure that the study is being conducted correctly. Both of these bodies ensure that all research is properly conducted and that the interests of those taking part are adequately protected.

What if I have any complaints about the study?

If you are unhappy about anything about the study and wish to complain, please contact the researcher in the first instance. You are also able to make a complaint through the normal NHS complaint procedure in NHS Ayrshire and Arran. Details about this are available through the research team. You can also submit a written complaint to the address below:

Complaints Team
PO Box 13, Eglinton House, Ailsa Hospital, Dalmellington Road, Ayr, KA6 6AB

Telephone: 01292 513620 **please note:** your call may be recorded
Fax 01292 513665
Email: complaintsteam@aapct.scot.nhs.uk

Appendix 2.8 Interview Schedule

Interview Schedule

1. **In relation specifically to the process of being diagnosed, what caused you the most distress?**
   - ➔ What were your initial thoughts and expectations?

2. **What has caused you the most distress in relation to living with MND?**
   - ➔ What changes have you found the most difficult?
   - ➔ How have you tried to cope with these?
   - ➔ What are the main differences between a good day and a bad day for you?

3. **Has having Motor Neurone Disease changed the way that you think or feel about yourself?**
   - ➔ Do you see yourself differently now than before you received your diagnosis?
   - ➔ How do you think other important people in your life see you?

4. **How have any roles you previously held changed?**
   - ➔ For example, in your relationships, working life or social activities.

5. **How well do you think that the distress thermometer and concerns checklist reflect areas which you have felt distressed (e.g. worried or upset) about?**
   - ➔ What areas do you think are missing, or what would you add?
   - ➔ How useful did you find it/do you think it would be in helping to guide a conversation with your clinician about what is important to you?
   - ➔ How much control do you feel that you have in appointments with clinicians?

6. **Do you think that services have helped you with the things that you most wanted help with? Has there been anything that you wanted help with that services did not help you with?**
   - ➔ Did you feel that your emotional needs and important areas in your life were discussed, as well as your physical health?
   - ➔ How would you describe your interactions with health care professionals?
   - ➔ What was helpful/unhelpful?

7. **What has your experience of uncertainty been like since your diagnosis?**
   - ➔ In your day to day life?
   - ➔ With regards to the future?

8. **Have there been any positive aspects of your experience that you would like to tell me about?**
   - ➔ Is there anything that we have not covered today that you'd like to discuss in relation to your experiences?

Appendix 2.9 Transcript Coding Example

Emergent Themes	Original Transcript	Exploratory Comments
Sense of control in knowing support is there	**: That's good, I can imagine that must make a difference.** P: There's someone on the end of the phone if I need them, eh… it makes me quite… quite happy, aye.	Support there when needed. Sense of control in being able to reach out for support and know he will receive this, predictable. Makes him happy
Wheelchair as uncertainty Loss of independence Focus on positives I'm not a worrier, focus on identity	**I: I'm glad of that. I was wondering what your experience of uncertainty had been like since your diagnosis?** P: Well the biggest uncertainty is… whether or when I'm going to end up in the wheelchair, eh, that's the biggest…biggest one of… is it tomorrow or…am I going to end up having a fall and that's me finished or… I mean, I started walking with basically, two walking sticks. But I kept falling and so I had to get this walker and I still fall now and again. But eh…I've no ended up in a wheelchair yet. No I cannae say there's any uncertainty other than that… eh, no, no…I tend not to worry about other things.	Wheelchair as uncertainty. Sense of mobility being important to sense of identity, maintain independence but there has been a gradual loss of this. Ultimate fear is losing this completely or having a fall and losing more mobility Having a fall as being finished as this would result in needing a wheelchair and not physically moving himself, others having to do this. When will it happen, *uncertainty reflected in how describing, jumping about in describing how this could happen.* Worry about falling. Decline of ability to walk. No other uncertainty *emphasised by repetition*. I don't worry about other things. I am not a worrier, not part of who I am.
I'm not a worrier Focus on identity Uncertainty difficult Signals giving up independence/control	**I: For you the uncertainty has, again, been focused on the wheelchair, and otherwise you've kept… managed to keep quite a positive attitude.** P: Aye, aye I…tend to do that, I don't let it get me down or worry me (mm hmm). The uncertainty of when it is going to happen is uh…the most, most sort of, concerning thing. I mean, it might be a while yet, I don't know when it will be, and that's…that's the thing.	*Repetition as emphasising positive attitude*. I don't get down/worried. I am not a worried or down person, restating identity and how this is out the norm for him. *Hesitant repetitive language reflecting this uncertainty.* Uncertainty of when he'll need a wheelchair as primary concern. Signals point where he'll give up some control and independence.
Hypervigilant to sings of feared things happening – maintaining control	**I: Definitely, that's the thing that might pop into your head and be a bit worrying.** P: Every now and again, is this the start of it, every time I get a wee stiffness or a wee nerve I go, oh… is this the start of it. That's it, that's mainly…what I think in my head.	A worry that's sparked by physical symptoms. Hypervigilant to signs of feared things happening. Trying to maintain what control he has, staying alert to be prepared.

Maintaining control Contentment over time End of the world initially Nothing has changed – identity wise Physical change acknowledge Alternative reasons – old age, covid Back to positive identity Retaining control	**I: That's understandable, you're almost looking out for the signs because it's so uncertain.** P: I look for the signs, definitely…so I can find out when it can happen. I mean it might be 3 years before it happens. **I: Do you think that experience of uncertainty has changed over time for you, because your diagnosis was so long ago?** P: Aye, it probably has, I'm probably more…more content with the fact I've got it (uh huh) ….initially it was…uh, It was the end of the world sort of thing… but as the years went on…but for years there's really been no change whatsoever, I'm just the same, nothing changed, all the same. But as I said, over the past couple of years… since the covid thing… we've all been less active, and I've been trying to move about more, maybe that's one of the reasons, I dunno. So…I've given up the [redacted]… that kept me active as well. And it's maybe age, old age…. I have quite a positive attitude and I don't…I don't let things get me down **I: That sounds like that's been a big factor in keeping you going.** P: Aye	Looking for the signs to find out when it happens. Realising it might not happen for some time. A strategy to maintain control. *Pause reflects reluctance around this being an inevitability.* Contentment over time. *Pauses reflecting trying to think about this experience in the past, a long time ago, becoming more fluid as it comes back to present do and how he feels now*. Initial thoughts of it being the end of the world, end of who he was as a person. No changes as meaning he is just the same as a person. Repetition of nothing's changed to emphasise this as being both no physically changes and no changes within himself. Reflecting on more recent changes, becoming more difficult to ignore. Attributing to potential alternative reasons, such as less activity during covid, old age. *Hesitations suggesting difficulty believing these other reasons – it's MND.* Returning to who he is – I am positive, I don't let things get me down and I am maintaining my identity and control by doing so.

Appendix 2.10 Sample of Reflective Notes

Reflection following interview with Marion

Prior to our interview, I had been feeling quite overwhelmed about the prospect of trying to meet multiple demands involving placement, research, and social commitments, most of which were being made easier by the ability to effectively communicate with others. When we began our interview, I was struck by how long it took for Marion to type the response that she wanted so that this could be heard through her verbal output device. I was very conscious of ensuring I allowed plenty of time for Marion to say everything she wanted for each question, and both of these factors lead to long silences between us both speaking. Marion told me about how she can feel like others do not always treat her as an equal, as they are not willing to give her the same opportunity to speak. I enjoyed speaking to Marion, but came off the call thinking about how infuriating I would find that experience, which is now her normality.

Marion had spoken about how she copes day to day, and had done this in such a poetic manner, despite her communication difficulties. She spoke about focusing on the small positives *("every day is a new day and the birds sing")*, which I found to be really powerful when we had just been discussing how much life has changed for her as a result of her MND diagnosis. Again, I had the experience of feeling both privileged to discuss such personal experiences with someone who had chosen to give up their time for my research, and struck by the resilience of the people I am interviewing.

It occurred to me that when Marion had been describing how she copes with her diagnosis and the ongoing uncertainty that this brings, she had been describing some principles of mindfulness. Reflecting back on previous interviews, similar principles had also been identified by other participants.

Appendix 2.11 Final Research Proposal

Name of Assessment: Major Research Project Proposal

Title of Project: Experiences of Distress and its Screening using the Distress Thermometer in Motor Neurone Disease

Name: Rachel King

Matriculation Number:

Academic Supervisor: Dr Jonathan Evans

Field Supervisor: Dr Luke Williams

Date of Submission: 09/06/2020

Version Number: 1

Word Count: 4744 including abstract and references (3327 without)

Experiences of Distress and its Screening using the Distress Thermometer in Motor Neurone Disease

Abstract

Background. The Distress Thermometer (DT) is a screening tool used to help identify distress across health conditions. Motor Neurone Disease (MND) covers a group of neurodegenerative diseases which primarily involve the progressive loss of motor function, but can also result in cognitive, behavioural and communication impairments . People with MND and their families face daily uncertainty due to the nature of these changes. Whilst the experience of psychological distress in people with MND is considered to be understandable it remains poorly understood and previous research has not examined how people with MND view the use of the DT as part of their assessment.

Aims. To better understand the lived experience of distress in the context of Motor Neurone Disease by exploring, i) participant experiences of distress within the context of a diagnosis of MND, if any; ii) the meaning of distress for individuals with MND; iii) participant's views of the Distress Thermometer and Concerns Checklist in relation to their own experiences.

Method. Semi-structured interviews will be conducted with people with MND. It is hoped that up to 6-8 participants will be recruited from the NHS Ayrshire & Arran MND Clinic. Interview transcripts will be analysed using Interpretative Phenomenological Analysis.

Applications. This research may help to inform understanding of distress within MND and in turn how this is assessed and incorporated into treatment plans across services.

Introduction

Psychological distress is commonly used as an indicator and outcome of mental health, but the concept remains unclear as it is applied to various symptoms, behaviours and difficulties (Drapeau et al., 2012). Distress has been defined in a healthcare context as ‘a multi-factorial construct involving a complex interplay between biological, psychological, interpersonal, social and existential factors’ with an emphasis on taking all of these aspects of distress into account (Brennan et al., 2012, p. 1346). It has been proposed that the term can encompass general worry, fears and sadness and spiritual or existential crises as well as mental health problems (Holland & Bultz, 2007). Measurement of distress has been recognised as important across different clinical populations and health care settings due to the negative impact it can having on treatment seeking and quality of life (Ransom et al., 2006; Donovan et al., 2014).

The Distress Thermometer (DT; Roth et al., 1998) and accompanying Concerns Checklist (CC) are tools which have been shown to be useful for screening distress and supporting health conversations across a range of health conditions in clinical practice (Snowden et al., 2011). The DT is a visual screening tool where individuals are asked to rate their level of distress over the past week on a scale from 0-10, with 0 indicating no distress and 10 indicating extreme distress. Alongside this, the CC allows patients to indicate the source(s) of their distress under various categories such as physical, practical and relationship problems. Brennan et al. (2012) have highlighted the importance of using the DT and CC as part of a structured, collaborative discussion in order to effectively identify and reduce distress in patients. Many studies attempt to establish the validity of the DT by comparing the DT to other screening measures. For example, various studies have established the validity of the DT as a tool to detect distress in cancer patients using this approach (e.g., Ma et al., 2014; Gessler et al., 2007). Evidence is more mixed for other health conditions and settings. Stewart-Knight et al. (2012) conducted a systematic review which found little evidence to support the validity of the DT across palliative care settings. However, they emphasised the importance of conducting qualitative interviews in order to understand experiences of distress, what factors contribute to distress and how this is reflected in the DT. The current study aims to understand whether people with MND feel that the DT and CC are useful in supporting health conversations and allowing them to identify their main concerns.

Health care conversations contribute to quality of patient care (Hemsley et al., 2015). It is particularly important to ensure that communication is effective when people experience communication difficulties as these individuals often do not receive support that they require to manage their difficulties (Wylie et al., 2013). One study found that both clients and professionals found preparation and structure helpful in reaching a shared understanding, which tools such as the DT and CC may assist with (Stans et al., 2018).

Motor Neurone Disease (MND) covers a group of neurodegenerative diseases which primarily involve the progressive deterioration of motor Neurones, which control voluntary muscle movement e.g., movement, speech, breathing and swallowing (Young et al., 2019). These impairments often lead to increased dependency, changes to lifestyle and relationships (Trail et al., 2009). Neuropsychological deficits affect over 50% of people with MND, and these can vary considerably. Both cognitive and behavioural impairments are negatively and significantly associated with disease stage (Crockford et al., 2018). Revised criteria for frontotemporal dysfunction MND have been expanded to incorporate the concept of a frontotemporal spectrum disorder to more accurately reflect changes in executive function, language, memory and behaviour which can occur in MND (Strong et al., 2017). Little is known about distress related to these changes as many studies focusing on wellbeing have excluded participants with cognitive or behavioural changes; however, behavioural changes have been linked to hopelessness in patients and to increased caregiver burden (Caga et al., 2019).

People with MND experience a wide range of psychological reactions, such as anxiety, hopelessness and distress (Averill et al., 2007). Depression and psychological distress have been found to be unrelated to disease progression or time since diagnosis (Rabkin et al., 2005). However, when psychological distress is present, it lowers quality of life (Johnston et al., 1999), which appears to be unrelated to functional impairment (Gibbons et al., 2013).

Psychological distress can also reduce survival rates and increase interest in hastened death (McDonald et al., 1994; Ganzini et al., 2002).

The experience of psychological distress in people with MND remains poorly understood. Research on the Distress Thermometer and Concerns Checklist has focused mainly on validating these against standardised diagnostic tools in various health conditions. However, measures examining distress often fail to address patients' holistic needs, instead focusing on depression and anxiety. It is important that distress is continually monitored in MND, as patients and their families face challenges in coping with uncertainty around the nature of changes on a daily basis. Needs change quickly with existing symptoms progressing as new ones develop, as patients continuously have to adapt to a decline in their abilities, an increase in their dependence on others and changes across all domains of their lives (Sakellariou et al., 2013). Bulbar symptoms lead to impairment of speech and affect up to 30% of people with ALS (Havervamp et al., 1995). The rate of speech impairment is difficult to predict and communication difficulties can be exacerbated by a loss of motor function. Communication difficulties change over time and have a significant impact on quality of life; it has been identified by MND patients as a key factor in maintaining or losing their independence (Gruis et al., 2011; Green et al., 2013).

The Motor Neurone Disease Clinic (MNDC) in NHS Ayrshire and Arran (NHS A&A) developed and introduced the DT with an adapted Concerns Checklist (MND CC) in 2017 to be administered routinely at clinic appointments. This tool allows patients to address physical, cognitive, emotional, practical and social needs as part of their care plan alongside clinicians, and ensures that clinicians enquire about distress. However, there are additional challenges to identifying psychological and emotional needs in the MND population due to communication difficulties and cognitive impairment, and its clinical utility has not yet been established. MND services who have audited the use of adapted versions of the DT and CC have found that these tools have allowed patients to identify unmet needs and prompted further referrals (Wright et al.,, 2011) and that these were acceptable to both patients and clinical staff (Lennox et al., 2018). However, it is worth noting that if there are no consequences of screening (e.g., lack of referral for an identified need), this may increase individual distress. A systematic review found that when screening identified distress in cancer patients and this was associated with a lack of follow up care, studies reported no benefit of screening, in contrast with studies which demonstrated improved patient outcomes when screening was linked with mandatory referral or intervention (Mitchell, 2013).

Aims

Research has suggested that individuals with MND are at risk of experiencing distress, in common with many other conditions. The Distress Thermometer and Concerns Checklist have been implemented within the NHS A&A MNDC to help identify signs and causes of distress, discuss these with clients and determine an appropriate intervention if necessary. However, views of individuals using this have not been fully explored. The proposed study aims to better understand the lived experience of, and the meanings associated with, distress in the context of MND. Interpretative Phenomenological Analysis (IPA) will be used to provide rich and detailed accounts of i) participant experiences of distress within the context of a diagnosis of MND, if any; ii) the meaning of distress for individuals with MND; iii) participant's views of using the Distress Thermometer and Concerns Checklist specifically in relation to their own experiences.

Plan of Investigation

Participants

Patients with MND will be recruited from the NHSA&A MNDC. All patients are given the adapted Distress Thermometer and Concerns Checklist at every clinic 15 minutes prior to their appointment and use this to help guide health conversations with professionals within the clinic. Clinics are not currently running routinely due to government restrictions on social distancing due to Covid-19. For those who have experience of using the DT/CC prior to these changes, their views of using the tools in relation to their own experiences will be explored. All participants will be sent out the tools along with participant information sheets prior to interviews. To what extent the DT and CC capture the experience of distress for participants will be explored.

Up to 6-8 participants will be recruited for this study.

Inclusion Criteria

Inclusion criteria: Diagnosis of Motor Neurone Disease; registered and seen within the Motor Neurone Disease Clinic; cognitive capacity to communicate informed consent; aged 18 years or older, ability to verbally communicate. Exclusion criteria: inability to communicate consent or lack of cognitive capacity to give consent as identified by the clinical team; communication difficulties which would be considered too difficult to understand via audio recording.

Recruitment Procedures

The researcher will provide information to MNDC staff about the study, its eligibility criteria and recruitment procedures in order to identify potential participants. The researcher will prepare a participant information sheet and opt-in form. Interested participants will be invited to send the opt-in form via post to the researcher or to complete this with a MNDC clinician to be passed onto the researcher. The researcher will then contact participants by telephone to arrange a time for interview at the participant's convenience and to allow an opportunity for further questions. Participants will be sent an outline of the project and interview process along with copies of the DT and CC. Concurrent data analysis will help to inform the recruitment process.

Interview

Semi-structured interviews will be conducted on an individual basis. Although interviews will be expected to last between approximately 1-1.5 hours, the researcher will be flexible with timings and number of interviews conducted. Participants will be given the option of multiple shorter interviews, and longer interviews will be conducted if required. In all cases, breaks will be taken as and when needed. Interviews will be recorded using digital recording equipment.

Field notes will also be taken by the researcher to supplement data. An interview schedule containing between 6 and 10 questions will be developed by the researcher, along with the study supervisor and MNDC staff. The interview schedule will also incorporate potential prompts to use during the interview process. Interpretative Phenomenological Analysis (IPA) aims to explore participants' experiences, understandings, perceptions and views, and assumes that participants seek to interpret their experiences into a form which is understandable to them.

Design

The design of the study will be a retrospective qualitative design with data being analysed using IPA. One to one interviews will be carried out with participants, and these interviews will then be transcribed and the content explored to identify recurring themes. This method of analysis will be used as IPA is concerned with how people make sense of their experiences and the process of the researcher making sense of this process in turn. This method will allow the research questions relating to people with MND's experiences of distress to be addressed appropriately.

Additionally, IPA places emphasis on researcher reflexivity, acknowledging that interpretations of data are tied to the researcher's ability to reflect and analyse. This is particularly important in this study as the researcher has no personal experience of MND, therefore engaging in reflective practice will allow the researcher to reduce the impact of preconceptions on interpretations.

Research Procedure

Interviews will be conducted by the researcher. Participants will be given three options of interview settings – in their home setting, via video conferencing software or at the MNDC - dependent on personal preference and government restrictions. Before commencing each interview, a brief outline of the research project and interview process will be given/sent to and read through with each participant. The DT and CC will be sent out prior to interview. The researcher will go over this with participants at the end of the interview and explore whether this tool has captured their experiences of distress or not. Confidentiality will be explained to each participant, including that identifying information will be removed and that pseudonyms will be used during the transcription of interviews and analysis. Permission will be sought for recording of the interview, and storage procedures will be explained. Participants will be reminded that their participation is voluntary and that they are free to withdraw at any time. Interviews will then be transcribed by the researcher and references to persons and places will be anonymised. Data analysis will commence concurrently with recruitment and data collection, which will help to inform when recruitment should be ceased (i.e., if saturation point is reached after fewer interviews). All interviews will be transcribed and checked by the researcher.

Data Analysis

Data will be analysed using Interpretative Phenomenological Analysis (IPA). Smith, Flowers and Larkin (2009) have set out the following 6 stages for analysis: (i) Reading and re-reading (ii) Initial noting (iii) Developing emergent themes (iv) Searching for connections across emergent themes (v) Moving to the next case (vi) Looking for patterns across cases. These steps will be followed when analysing data in this study.

Justification of Sample Size

The method of IPA is typically based on, and is best suited to, small sample sizes involving individual interviews of participants. . In IPA research purposive sampling is normally used in order to have a homogenous sample. This approach will be used as much as possible but will be dependent on respondent rates and by the small population of people with MND. For doctoral level research projects, the recommended sample size is 6-8 participants (Turpin et al., 1997). Therefore, the study will seek a sample size of up to 6-8 participants, although this number will be informed by concurrent data collection in terms of when the point of saturation is reached with regards to richness of themes and topics explored.

Settings and Equipment

Participants will be given three options of interview setting depending on preferences and government restrictions: 1) the MNDC, where interviews will be conducted face to face and recorded with an audio recorder. In this instance, interviews will be conducted within office hours (9-5) in a clinic room. Interviews will be conducted at times when other clinicians are within the clinic space and notified that interviews are taking place. 2) participant’s homes. In this setting, Lone Working policies for both NHS A&A and the University of Glasgow will be followed, and the researcher will keep these interviews within MNDC clinic hours. 3) NHS A&A approved video conferencing software such as Attend Anywhere. This option may be more convenient for participants and may be necessary as part of government response to Covid-19. Software such as Skype has been used in previous studies utilising this method (e.g., Ferolino et al., 2020) and is suggested as the most feasible alternative to interviews conducted face to face (Hanna, 2012). In this instance, NHS A&A guidelines regarding remote appointments will be followed by the researcher. Participants will be able to access a waiting room via a link which will be sent by letter or email after confirming the time of the interview that suits the participant.

In all settings, participants will have the opportunity to reflect on how they are feeling at the end of the interview and to contact a psychologist or nurse from the MNDC. Participant information sheets, consent forms, copies of the DT/CC and stamped addressed envelopes will be required for participant recruitment. If participants are unable to sign a consent form due to limb weakness, they will be invited to give verbal consent in the presence of a witness (e.g., caregiver or MNDC staff member) and the researcher will record this in writing and using a voice recorder.

Interviews will be conducted on a one-to-one basis and will be audio-taped using a voice recorder. Recordings will be transferred to and stored on a University of Glasgow encrypted laptop, with recordings being deleted from the recording device. Transcriptions will be anonymised and stored on the laptop. Back up copies of both recordings and transcripts will be stored within the Glasgow University Network, in a password protected folder only accessible to the researcher and supervisor. When the study is completed, recordings will be deleted from the university network. Transcripts will be retained on enlighten for a period of 10 years in keeping with policy at the University of Glasgow and will be deleted after this period.

Health and Safety Issues

Researcher Safety Issues

Where possible, interviews will take place within normal working hours in MNDC office and will comply with standard safety procedures. MNDC staff will be aware of when interviews are being conducted and will be available if required. Home visits will be conducted as necessary and both the Lone Working Policy provided by NHS A&A and the Lone Study Procedure provided by the University of Glasgow will be followed.

Participant Safety Issues

Written or verbal consent will be obtained from the participant and the participant information sheet will detail information on the opportunity to opt-out at any time. The researcher will explain to participants that they can stop the interview or take a break at any time and will regularly check in with clients to check for fatigue or distress. If participants disclose risk of harm to self or others, the researcher will act appropriately while respecting limits of confidentiality.

Ethical Issues

All information gathered during participant interviews will remain confidential and will be stored in a locked filing cabinet within NHS A&A, and will be transferred to a locked filing cabinet within the University of Glasgow upon study completion. An NHS Research Ethics Committee and the Research and Development Department in NHS A&A will be contacted for approval of the proposed research.

Due to the nature of the topic, it is expected that participants may become upset or distressed during or following interviews. During interviews, the researcher will frequently check in with participants to check for fatigue or distress and ensure they are happy to continue. The researcher will allow time at the end of interviews for participants to reflect on how they are feeling. All participants will have the opportunity to contact a nurse or psychologist from the MNDC for support after their interview and relevant clinicians will be aware of when interviews are taking place.

Financial Issues

Funding will be required for printing up to 20 participant information sheets, up to 10 consent forms, up to 10 copies of the DT/CC and up to 20 stamped addressed envelopes for interested participants. A digital voice recorder and transcribing kit will be borrowed from the University of Glasgow.

Timetable

Submit draft proposal: January 2020

Application to ethics and R&D: May 2020

Recruitment commencement: October 2020

Data analysis: October 2020-May 2021 (concurrently with recruitment)

Writing-up: June-July 2021

Practical Applications

The results of the proposed research will be written up as part of a Clinical Psychology doctoral thesis and will be published on the Glasgow University thesis website. The research will be shared with interested participants, MNDC staff and submitted for publication. It may inform understanding of distress within MND and in turn how this is assessed and incorporated into treatment plans. The current research would help to provide insight into how the adapted DT and CC are experienced by people with MND in relation to their own experiences of distress.

Ideally the current study would seek to include the experiences of people who have significant communication difficulties, particularly given the additional distress that these impairments are known to bring. However, this is out with the scope of this research and would require ethical considerations with regards to putting people with significant communication difficulties through the process of taking part in lengthy interviews. Further research should seek to better understand the experience of distress of this population.

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Appendix 2.12 COREQ Checklist for this Study

COREQ (COnsolidated criteria for REporting Qualitative research) Checklist

A checklist of items that should be included in reports of qualitative research. You must report the page number in your manuscript where you consider each of the items listed in this checklist. If you have not included this information, either revise your manuscript accordingly before submitting or note N/A.

Topic	Item No.	Guide Questions/Description	Reported on Page No.
Domain 1: Research team and reflexivity			
Personal characteristics			
Interviewer/facilitator	1	Which author/s conducted the interview or focus group?	55
Credentials	2	What were the researcher's credentials? E.g. PhD, MD	1
Occupation	3	What was their occupation at the time of the study?	56
Gender	4	Was the researcher male or female?	56
Experience and training	5	What experience or training did the researcher have?	56
Relationship with participants			
Relationship established	6	Was a relationship established prior to study commencement?	54
Participant knowledge of the interviewer	7	What did the participants know about the researcher? e.g. personal goals, reasons for doing the research	54
Interviewer characteristics	8	What characteristics were reported about the inter viewer/facilitator? e.g. Bias, assumptions, reasons and interests in the research topic	
Domain 2: Study design			
Theoretical framework			
Methodological orientation and Theory	9	What methodological orientation was stated to underpin the study? e.g. grounded theory, discourse analysis, ethnography, phenomenology, content analysis	55/56
Participant selection			
Sampling	10	How were participants selected? e.g. purposive, convenience, consecutive, snowball	54/55
Method of approach	11	How were participants approached? e.g. face-to-face, telephone, mail, email	54
Sample size	12	How many participants were in the study?	54/55
Non-participation	13	How many people refused to participate or dropped out? Reasons?	54/55
Setting			
Setting of data collection	14	Where was the data collected? e.g. home, clinic, workplace	55
Presence of non-participants	15	Was anyone else present besides the participants and researchers?	55
Description of sample	16	What are the important characteristics of the sample? e.g. demographic data, date	57
Data collection			
Interview guide	17	Were questions, prompts, guides provided by the authors? Was it pilot tested?	127-128
Repeat interviews	18	Were repeat inter views carried out? If yes, how many?	55
Audio/visual recording	19	Did the research use audio or visual recording to collect the data?	55
Field notes	20	Were field notes made during and/or after the inter view or focus group?	56
Duration	21	What was the duration of the inter views or focus group?	55
Data saturation	22	Was data saturation discussed?	55
Transcripts returned	23	Were transcripts returned to participants for comment and/or	

Topic	Item No.	Guide Questions/Description	Reported on Page No.
		correction?	
Domain 3: analysis and findings			
Data analysis			
Number of data coders	24	How many data coders coded the data?	56
Description of the coding tree	25	Did authors provide a description of the coding tree?	
Derivation of themes	26	Were themes identified in advance or derived from the data?	56
Software	27	What software, if applicable, was used to manage the data?	
Participant checking	28	Did participants provide feedback on the findings?	
Reporting			
Quotations presented	29	Were participant quotations presented to illustrate the themes/findings? Was each quotation identified? e.g. participant number	57-70
Data and findings consistent	30	Was there consistency between the data presented and the findings?	57-70
Clarity of major themes	31	Were major themes clearly presented in the findings?	57-70
Clarity of minor themes	32	Is there a description of diverse cases or discussion of minor themes?	

Developed from: Tong A, Sainsbury P, Craig J. Consolidated criteria for reporting qualitative research (COREQ): a 32-item checklist for interviews and focus groups. *International Journal for Quality in Health Care*. 2007. Volume 19, Number 6: pp. 349 – 357

Once you have completed this checklist, please save a copy and upload it as part of your submission. DO NOT include this checklist as part of the main manuscript document. It must be uploaded as a separate file.