Flinders University

Exploring the interaction between cognitive and behavioural changes and dysphagia in Motor Neurone Disease management.

By

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ABSTRACT

Introduction

The interaction between dysphagia (impaired swallowing) and cognitive and behavioural changes in Motor Neurone Disease (MND) has not yet been explored. This interaction is important as almost all people with MND (pwMND) will develop dysphagia over disease progression. Additionally, MND is a multisystem disease with cognitive and behavioural changes occurring in up to 50% of pwMND. Cognitive and behavioural changes in MND are linked to negative outcomes, including shorter survival times, impaired health care decision-making, reduced quality of life and increased caregiver burden. However, it is currently not yet understood how cognitive and behavioural changes in MND interact with dysphagia management.

Consequently, this program of research makes a novel contribution to knowledge in that it raises awareness of the interaction between cognitive and behavioural changes and dysphagia in MND. This research begins to shed light on some of the negative consequences that may occur for families living with MND, who are managing dysphagia in the presence of co-occurring cognitive and behavioural changes. The research question underpinning this program of research is: *How do cognitive and behavioural changes in MND interact with dysphagia management?*

Methods

Grounded in constructivism and interpretivism, this thesis consists of five qualitative studies and one mixed methods study. Initially, two scoping literature reviews were conducted to synthesise the current literature. These investigate the interaction between dysphagia and cognitive and behavioural changes, and the management of cognitive and behavioural changes more broadly. An ethnographic study was planned, however, due to Covid-19 restrictions, was not able to be completed. Consequently, a mixed methods survey was

conducted with health care professionals (HCPs) to identify their current clinical practices related to cognitive and behavioural changes in MND. Following this, semi-structured interviews were undertaken in two separate research projects. The first study built on from the data identified in the survey and investigated the experiences and attitudes of HCPs who support families living with MND. The second study investigated the experiences and preferences of families living with MND regarding both dysphagia and cognitive and behavioural changes. The revised Theoretical Domains Framework was used to deductively analyse these data and to identify barriers and facilitators to practice. Secondly, thematic analysis was conducted to inductively identify themes related to these data.

Results

The key findings of this program of research included: i. The relationship between cognitive and behavioural changes in MND and dysphagia has not been explicitly investigated in the existing literature (Chapter 3) and ii. the information to guide practice related to managing cognitive and behavioural changes in MND more broadly is lacking (Chapter 5). iii. HCPs identified that managing cognitive and behavioural changes in MND is important, however described knowledge gaps and resource burdens that impact on their ability to manage this (Chapter 6 and 7). iv. Further, families living with MND described increase burden and distress associated with cognitive and behavioural changes in MND, and believed it was an important area of MND to learn about from their HCPs (Chapter 8).

Conclusion

This program of research demonstrates the complexities for pwMND and their carers as they navigate cognitive and behavioural changes in the presence of dysphagia. This research shows that cognitive and behavioural changes impact on the quality of life of families living with MND and contributes to relationship breakdown and increased stress for both pwMND and carers. Furthermore, HCPs caring for families living with MND are not trained in cognitive and behavioural changes associated with MND and there is limited MND-specific

guidance available to support them. More work in needed to investigate this phenomenon to explore families' preferences for receiving information about and management of cognitive and behavioural changes in MND.

DECLARATION

I certify that this thesis:

1. does not incorporate without acknowledgment any material previously submitted for a degree or diploma in any university

2. and the research within will not be submitted for any other future degree or diploma without the permission of Flinders University; and

3. to the best of my knowledge and belief, does not contain any material previously published or written by another person except where due reference is made in the text.

Signed

Date 23/10/2024

ACKNOWLEDGEMENTS

I would like to express my deepest gratitude to all the participant's invaluable contributions that made this research possible. Specifically, to the HCPs who graciously reflected on their clinical practice, and candidly shared their experiences, I thank you. The care and concern you have for the people you support is clear. Your willingness to share your time and insights provided rich context for this research, and your input has been essential in advancing our understanding of cognitive and behavioural changes in MND. Without your openness and cooperation, this work could not have been completed. Thank you for supporting me, trusting me, and for allowing me the privilege of learning from you.

To the pwMND and carers, I am eternally grateful. I am deeply touched by your stories, and I hope that this body of work accurately represents your experiences and begins the work that is needed to improve aspects of MND care. Throughout our conversations, you expressed an awareness that contributing to this research may not help you directly, but your hope was that you may in some way help others experiencing this disease. I deeply respect this generous and selfless view.

I would like to also extend my heartfelt thanks to my supervisors, Professor Sebastian Doeltgen and Associate Professor Stacie Attrill, for their guidance, encouragement, and wisdom throughout this research journey. Your expertise, patience, and insightful feedback have been invaluable in shaping this thesis. I am deeply grateful for your mentorship, for believing in me, and for always pushing me to strive for excellence. Thank you for your time, support, and the many discussions that helped me to navigate the challenges and grow as a researcher. Stacie, I am in awe of how your mind works! Seb, you have such a kind and gentle soul. You have managed to ground me and calm me when I was feeling overwhelmed and frustrated. Thank you for the coffee catchups and many lengthy debriefs!

Lastly, but certainly not least, to my family. Keira and Reed, my hope is that one day you will look back on my journey and see how far, as a family, we have come. You already have some insight into the beginnings of my life. A childhood that was filled with challenges, growing up with little and facing much adversity. A little girl, who never dreamed that university was possible. My wish is that by witnessing me take on this challenge, you will be inspired to pursue your own dreams, no matter how much of a challenge they may seem. I hope you know that while the path may not always be easy, with determination, you have what you need to make it possible. Always strive to be the best version of yourselves. I love you both, more than you will ever know.

To my partner Troy, your unwavering support and understanding throughout this journey has been incredible. You have encouraged me during the difficult moments when I wanted to throw in the towel (and brought me a thousand of cups of tea!). When I was sitting in a room of darkness, you could see the light at the end of the tunnel. Often, you believed in me way more than I could. This helped me to forge ahead. I am grateful for having you by my side. I love you.

PUBLICATIONS AND PRESENTATIONS FROM THIS DOCTORATE

PUBLICATIONS:

The following publications reflect the dissemination to date of some of the research conducted as part of this program of research.

1. **Francis, R.,** Attrill, S., & Doeltgen, S. (2021). The impact of cognitive decline in amyotrophic lateral sclerosis on swallowing. A scoping review. *International Journal of Speech-Language Pathology*, *23*(6), 604–613. https://doi.org/10.1080/17549507.2021.1894235

This paper relates to the data presented in Chapter 3.

2. **Francis, R**., Attrill, S., Radakovic, R., & Doeltgen, S. (2023). Exploring clinical management of cognitive and behavioural deficits in MND. A scoping review. *Patient Education and Counseling*, *116*, 107942. https://doi.org/10.1016/j.pec.2023.107942

This paper relates to the data presented in Chapter 5.

Additional publications that are not part of this thesis, but related to this body of research:

1. Paterson, M., Doeltgen, S., & **Francis, R.** (2024). Sensory Changes Related to Swallowing in Motor Neurone Disease. *Dysphagia*, 1-12. https://doi.org/10.1007/s00455-024-10742-x

ORAL PRESENTATIONS:

1. **Francis, R.,** Radakovic, R., Attrill, S., & Doeltgen, S. *How is Cognitive and Behavioural Change in MND Managed?* Australia and New Zealand MND Research Symposium. Platform presentation, November 2023. (Chapter 5)

2. **Francis, R.,** Attrill, S., & Doeltgen, S. *How do health care practitioners recognise, provide information on, and manage cognitive and behavioural symptoms associated with MND? Exploring practices, enablers and barriers*. Australia and New Zealand MND Research Symposium. Platform presentation, August 2024. (Chapter 6)

3. **Francis, R**. '*State of Play' webinar series*. MND Research Australia. Online presentation, October 2024

CONFERENCE PRESENTATIONS - POSTER:

1. **Francis, R.,** Attrill, S., & Doeltgen, S. *Interaction between decline of swallowing and cognitive function in MND*. 30th International Symposium on ALS/MND 2019, Perth. Poster presentation, December 2019.
2. **Francis, R.,** Attrill, S., & Doeltgen, S. *The impact of cognitive decline in ALS on swallowing is not understood or researched*. Dysphagia Research Society. Online poster presentation, June 2020

AWARDS

This award was received to support travel costs for a poster presentation for the data presented in Chapter 3

1. Nina Buscombe Travel Award ($500) for travel to the MND International Symposium, 2019, Perth, WA.

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Glossary

Carer	A person who cares for someone and provides day-to-day living support. This may be a spouse, family member, or paid support worker.
Discipline	A branch of health care knowledge, which requires formal education, training and a variety of specialised skills relevant to a specific scope of practice. Health disciplines apply scientific principles and evidence-based practice to optimise patient/client outcomes.
HCPs	Formally qualified health care professionals e.g., medical doctors, allied health professionals, nurses or clinical support people who work to support people manage a disease or illness.
Management	An organised, proactive, multi-component approach to healthcare delivery that involves all members of a defined population who have a specific disease entity. Care is focused on and integrated across the entire spectrum of the disease and its complications, the prevention of comorbid conditions, and relevant aspects of the delivery system. Essential components include identification of the population, implementation of clinical practice guidelines or other decision-making tools, implementation of additional patient, provider, or healthcare system-focused interventions, the use of clinical information systems, and the measurement and management of outcomes Definition adapted from Norris et al. (2003)
Percutaneous endoscopic gastrostomy	A method of inserting a feeding tube used in gastrostomy. The tube is inserted directly into the stomach via a small incision in the abdominal wall guided via endoscope.

Note to the reader:

The spellings ‘Motor Neurone Disease’ and ‘Motor Neuron Disease’ are both used within the literature. ‘Motor Neurone Disease’, will be used within this thesis as this is the term used by both the Motor Neurone Disease Association of Australia and the Motor Neurone Disease Association of the United Kingdom. Further, Australian English spelling is most similar to British English.

The term ‘Motor Neurone Disease’ is used predominately within Australia, the United Kingdom and New Zealand. Within the United States however, MND is referred to as Amyotrophic Lateral Sclerosis (ALS). Within this thesis, Motor Neurone Disease (MND) will be used throughout, however note the term ALS was used in the published articles shown in Chapter 3 and Chapter 5.

Abbreviations and commonly used acronyms within this thesis

AAC	Augmentative and alternative communication
AHP	Allied Health professional
ALS	Amyotrophic lateral sclerosis
ALS-FRS	Amyotrophic Lateral Sclerosis Functional Rating Scale
BMI	Body mass index
CPGs	Central pattern generators
CNI	Cranial Nerve One Olfactory
CNII	Cranial Nerve Two Optic
CNIII	Cranial Nerve Three Oculomotor
CNIV	Cranial Nerve Four Trochlear
CNV	Cranial Nerve Five Trigeminal
CNVI	Cranial Nerve Six Abducens
CNVII	Cranial Nerve Seven Facial
CNVIII	Cranial Nerve Eight Vestibulocochlear
CNIX	Cranial Nerve Nine Glossopharyngeal
CNX	Cranial Nerve Ten Vagus
CNXI	Cranial Nerve Eleven Accessory
CNXII	Cranial Nerve Twelve Hypoglossal
coMND	Carer of MND
CSF	Cerebral spinal fluid
DNA	Deoxyribonucleic acid
DT	Dietician
ECAS	Edinburgh Cognitive & Behavioural ALS Screen
FA	Flail arm syndrome
FL	Flail leg syndrome
FDA	American Food and Drug Administration board
FTD	Fronto-temporal dementia
GP	General medical doctor
HCP	Health care professional
HCPs	Health care professionals
HREC	Human research ethics committee
LMN	Lower motor neurone

MCI	Mild cognitive impairment
MD	Medical doctor
MDT	Multidisciplinary team
MND	Motor neurone disease
MND/FTD	Motor neurone disease frontotemporal dementia continuum
NDIS	National Disability Insurance Scheme (Australia)
NFs	Neurofilaments
NfL	Neurofilament light chains
NHS	National Health Service (UK)
PBA	Pseudobulbar affect
PBP	Pseudo-bulbar palsy
PICF	Participant Information and Consent Form
PLS	Primary lateral sclerosis
PMA	Progressive muscular atrophy
PMC	Primary motor cortex
pwMND	People or person with Motor Neurone Disease. Used in both singular and plural forms as appropriate in the context of the sentence.
RNA	Ribonucleic acid
SOD1	Superoxide dismutase 1 gene
SSRIs	Selective serotonin reuptake inhibitors
TDF	Theoretical Domains Framework
TDP-43	TAR DNA-binding protein 43
TGA	Therapeutic Goods Administration
ToM	Theory of Mind
UMN	Upper motor neurones (Upper motor neurons)
UOS	Upper oesophageal sphincter

Note to the reader:

Throughout this thesis the following acronyms and terms will be frequently used. A descriptor of these is provided in Table 1:

Table 1

Acronyms with descriptor frequently used within this thesis

Acronym or term	Descriptor
Carer	The term carer will be used to represent the pwMND's closest family member, which is typically a spouse, however, may also refer to a professional support worker who is caring for a pwMND as their primary carer.
coMND	This acronym is used in Chapter 8 as a participant code. It represents participants who are a carer of a person with MND.
Families with MND	The term families with MND will be used to describe the wider family unit, of which one member is a pwMND.
HCP/HCPs	A heath care professional, from any medical or allied health field, who advises on or applies preventive and curative measures, and promotes health to meet the health needs and expectations of families living with MND. When used as a possessive, the acronym will be written as HCPs'.
MND	Motor neurone disease
pwMND	An individual person who has been diagnosed with MND. This acronym is used in both singular and plural forms as appropriate in the context of the sentence. When used as a possessive, the acronym will be written as pwMND's

REFLEXIVE STATEMENT

It is good to have an end to journey towards; but it is the journey that matters, in the end.
Ursula K. Le Guin. The Left Hand of Darkness, Ch. 15, 1969

This PhD has ended up very differently to how it began.

The pathway to my PhD started in 2018, following on from the completion of a Bachelor of Speech Pathology, Honours degree. My Honours project was situated in quantitative research and involved the use of the innovative technology of pharyngeal high-resolution impedance manometry. Here, I investigated the effect of volume variance on pharyngeal swallowing biomechanics, in healthy older people. The goal of this project was to better understand the physiology of swallowing function in relation to bolus movement, and specifically, to understand what effect variances to bolus volume had on biomechanical action. This project was conducted, as a better understanding of swallowing biomechanics in healthy swallowing may help to inform speech pathologists working with people who have impaired swallowing function, known as dysphagia.

After successfully completing Honours with a First Class (90) result, it felt natural to move from that completed project, which I greatly enjoyed and learnt so much from, straight into a PhD, to build upon and further develop my newly acquired research skills. With encouragement from my supervisor, I decided to embark on a program of research, which would investigate impaired swallowing function in motor neurone disease. For a visual representation of the timeline of my PhD, please refer to Figure 1.

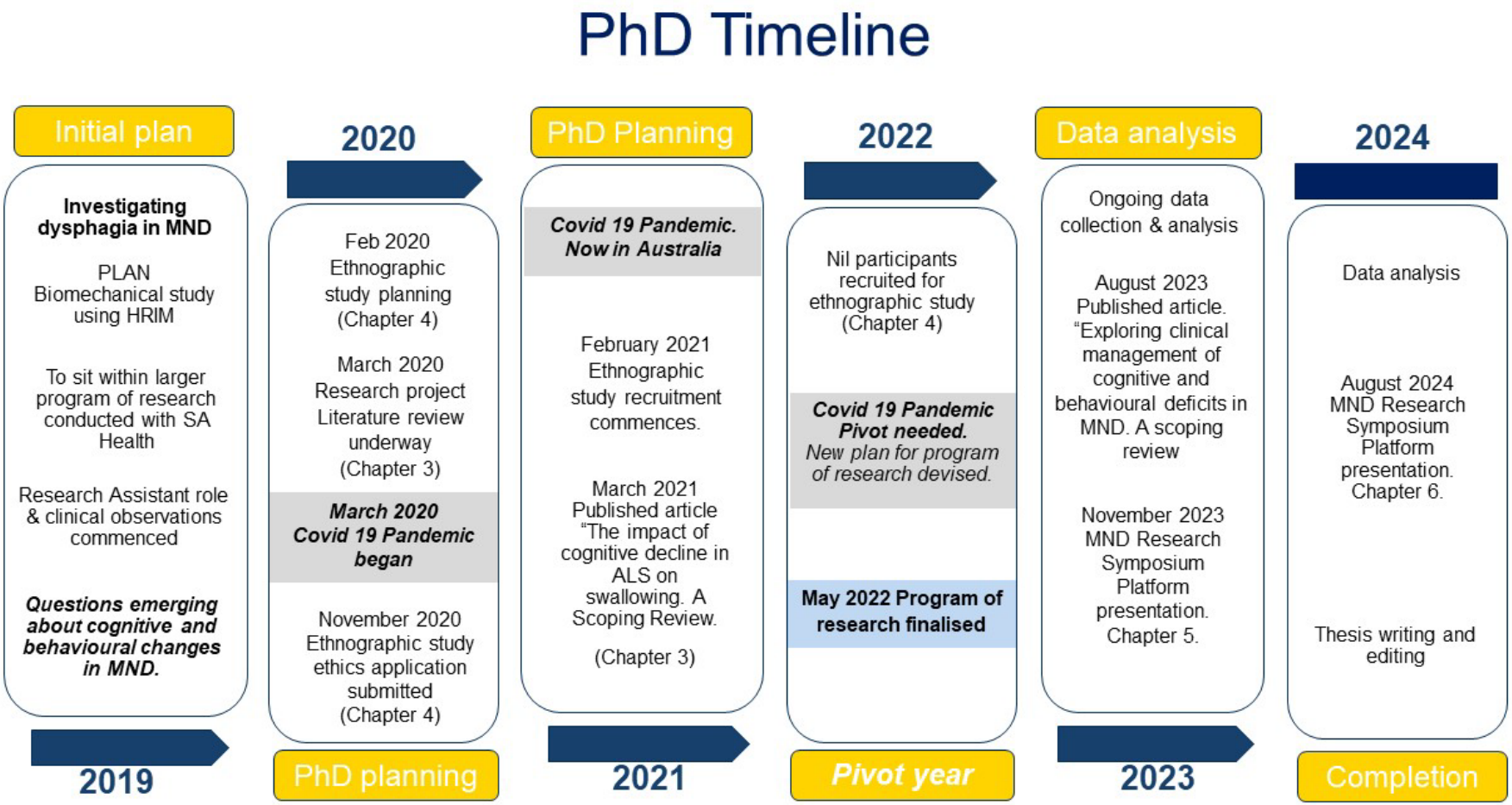

Figure 1: Doctor of Philosophy (PhD) program of research timeline. Image by thesis author.

The aim of my Doctor of Philosophy (PhD), at its inception, was to better understand how the biomechanics of swallowing are impacted in pwMND, using high-resolution impedance manometry. I was excited by the prospect of embarking on a research project that had the potential to inform understanding of swallowing decline in MND. The benefits of this knowledge had the potential to inform clinical practice and improve how speech pathologists provide care for pwMND, who are experiencing dysphagia.

Through intensively studying the existing literature, my understanding of the variability of MND deepened, and I began to realise that the complex disease mechanisms, genetics factors and varying phenotypes meant that no two pwMND experienced the disease in the same way. For some, they were confronted with rapidly progressing symptoms and a life expectancy of only two and a half-year's survival post the onset of symptoms. Whilst for others, the disease progressed slowly, taking away motor function initially in only one area of their body, but greatly impacting on their quality of life. Regardless of these variations, there is no known cause, and no known cure for MND. In Chapter 1, I expand on my learnings related to MND, with a detailed description of the current state of knowledge of the disease.

As a speech pathologist, I was aware of my role in supporting people who are experiencing dysphagia. Speech pathologists play an important role in supporting pwMND, as almost all people with the disease will eventually develop dysphagia as the disease progresses. I studied the literature, then realised I wanted to find out more about the clinical management of MND, so in the very early stages of my PhD journey (in 2019) I participated in observational opportunities in clinical settings.. This, along with my continued reading allowed me to develop expert knowledge of MND, as well as an increased awareness of the functional and psychological impacts of the disease on families. This experience was a pivotal moment, which shaped my program of research. I was able to better understand the role of a speech pathologist who supports pwMND to continue to eat and drink safely, to manage their secretions and to decide when (or if) it is the right time to move to gastro-

enteral feeding.

I had the opportunity to meet many pwMND and I observed first-hand the variability of this disease. I was beginning to better understand the phenotypes and how this meant that no two people had the same experience when it came to an MND diagnosis. The story of one family was particularly emotional, not just for me, but for all HCPs involved with their care. This person was young and active, they were a partner, a parent, and a friend to many. I was there the day they received their diagnosis of bulbar onset MND. It was an emotional day for all.

In just four short months, I observed the disease progress right before my eyes. They quickly became anarthric and were no longer able to walk. They died in less than a year. The parallels between this person's life and my own made this loss even harder to process. In contrast, another person I met in the clinic had a slow progressing type of MND, which only impacted on the function of their lower motor neurones. A rarer case, they had survived for over fifteen years. They were no longer able to move any part of their body, but still had the ability to talk and eat. I listened as they told me how terrified they were of choking. Their ability to cough had now been taken away by the disease, and they were frightened that no one would notice as they weren't able to gesture to get help. I found myself thinking deeply about quantity vs. quality of life.

As I continued to read and continued to observe, I also learned that MND is associated with cognitive and behavioural changes in some people with the disease. This was a surprise to me, as even the name suggests that this disease purely impacts on motor neurones and that cognitive function is spared. From further reading, it became clearer that this knowledge was slowly emerging in the literature, but not everyone working in the space, or living with the disease, was aware of this symptom.

I witnessed first-hand the rapid deterioration of a bulbar onset type MND, with people

experiencing severe functional changes to their speech and swallowing. At the same time, I was also observing parallels between what I was seeing in people with MND and what I was reading in the published literature. Bulbar onset MND typically progresses more rapidly and is more likely to be associated with cognitive and behavioural changes. I participated in several discussions with HCPs around this topic, and they too expressed worry that some people they were supporting were experiencing cognitive and behavioural changes. Through observation and literature exploration I noticed that there appeared to be no formal clinical pathways or guiding frameworks established to assess or manage cognitive and behavioural changes.

This is when the pathway of my PhD began to change.

In the initial research project, we were proposing three monthly swallowing studies, meaning participants (with MND) would need to attend a data collection session, for a minimum of an hour and undergo an invasive transnasal scoping procedure. Added to this, was travel, which may exacerbate feelings of fatigue that are associated with MND. I began to reflect on the ethics of conducting invasive swallowing research on this population and found myself questioning if the benefits of this research would create an undue burden for the participants.

As outlined in the Good Practice Guidelines for research (National Health and Medical Research Council, 2023), any anticipated benefits of research must outweigh any foreseeable risks. I wondered if conducting invasive swallowing studies with pwMND would benefit this population, given the terminal prognosis. I was not sure if the benefits of understanding swallowing biomechanics outweighed the burden that I would be adding to pwMND, through repeated, unpleasant transnasal catheter assessment procedures. In addition, asking people to travel for research, given impaired mobility and fatigue related to MND, was a conflicting factor I began to consider. At the same time, I also found my focus was shifting to other characteristics of the disease I was observing and reading about, such as cognitive and behavioural changes. I observed, some pwMND respond to care

recommendations with reluctance or refusal, whilst their carer was encouraging them. I questioned if this refusal was because they truly did not want the intervention, or if perhaps, this was because they were having difficulty understanding the new and complex information. As I observed, I reflected upon my readings into MND being associated with apathy, rigid thinking and impaired decision-making.

As I met more pwMND, I wondered often, if someone is unable to speak, and their motor function is also impaired, how are HCPs able to determine the person's ability to comprehend the information about treatment options and management of MND? Clinical consultations often occur with a person, who was now no longer able to move or speak. Conversations about their care seemed to be happening around them. It was not clear to me how much they understood about the changes they were experiencing, or if they were able to consider the options being presented to them. Clinical conversations were often had between the HCP and the carer, and management decisions were often left for the carer to make. Some excerpts from my observational diary reflect these thoughts:

Observational diary entry 11/4/2019:

> Continued thoughts around cognition occur for me as I observe. Neurological vs psychological factors of MND such an interesting area to discover. Have observed people that are very proactive in their care, reading, researching and wanting PEGs fitted ASAP and alternatively observed people that are wanting to wait, see what happens. Have also observed almost childlike presentation in some patients e.g. "I think I'm doing a terrific job with my swallowing" 70yr old male with MND.
>
> "Did I beat my IOPI tongue strength test score from last time?" 31yr old male with MND. Laughing at the OME instructions. Is this how he was premorbid? Is it

> emotional lability/cognitive decline? Is it denial of disease? So interesting to test for psych/cog (FBA?) and have a carer's perspective on what they think the behavior changes are like. Literature suggests that cog changes can occur before the person has any reason to go to DR with limb/speech/swallowing symptoms.

As I continued to read and continued to learn about cognitive and behavioural changes in MND, and as I met more families impacted by this disease, it made we wonder how we could improve care. Why were cognitive and behavioural changes in MND not consistently being talked about? Why was screening not being conducted as usual practice? How can HCPs be certain that the pwMND fully understands the information being explained to them? How could a HCP be certain that the pwMND was making decisions with capacity?

Similarly, the dysphagia management strategies that are recommended by speech pathologists require cognitive engagement, to understand and enact. These management strategies include postural adaptations, diet modifications and swallowing manoeuvres. I reflected that the broader community rarely consider their swallowing function. Swallowing is something that humans do without thinking, just like breathing. So, to have to implement changes to this, requires a person to have awareness and insight. I increasingly suspected, given the literature that I was reading, that awareness and insight had the potential to become impaired in people with cognitive and behavioural changes in MND:
Observational diary entry 26/6/2019:

> Sat in with a HCP as they explained feeding via a PEG to a patient who had just given consent to the procedure.. The whole thing felt a bit strange, and I was surprised by that. I watched the two elderly people trying to understand the information that they were being given and trying to navigate all this tricky and confronting news and it seemed that they were quite baffled and overwhelmed. The female patient was severely dysarthric so she could not participate in the

conversation about her disease progression. It often seems that healthcare professionals lead the conversation, with patients having fewer chances to ask questions or ensure they fully understand the information being shared. Given that cognition and behavior change can occur in 50% of pwMND I feel that language needs to be simplified and supported with visuals, or information sheets. There is some jargon used and most speak at a very fast rate etc.

I would love to see more written information given out with strategies and recommendations. And visuals used to support the information that the clients are being given. Show them what a PEG looks like as the procedure is explained.

Often after these observations, I would leave feeling terribly sad. I reflect on the time before I became a HCP, and realise that as a lay person, I had a somewhat naïve view of medicine, believing that HCPs would always know the right thing to do and say for any medical presentation. I wonder if pwMND and their families hold the same view. I reflect on how confronting it could be to learn that, not only do you have a terminal disease, but also there may not be any answers to solve your clinical problem.

From my clinical observations, I was beginning to see that HCPs invest themselves into caring for their patients, but are often restricted by time, availability of clinical resources and policy:

Observational diary entry 21/11/2019

It has occurred to me how difficult it is to pick up on cog decline in people who have MND as they also have reduced ability to talk, reduced ability to gesture, reduced facial expression and reduced movements so it is so hard to others to know what is going on with their frontal lobe!

I did a screening assessment with a lady who has bulbar onset MND, she is severely dysphagic, NBM with all nutrition via PEG, anarthric and uses an AAC to communicate (writing board) so we did the ALS CBS in written form.

Her husband made a comment that she has “not lost her marbles”, however her ALS CBS score was only 9 (out of 20). Scores below 10 indicate a possible FLTD and require further assessment. I observed her while doing the screener it seemed obvious to me that she had difficulty understanding the instruction – particularly when writing the months of the year backwards starting at December. In her response she wrote the word December and then proceeded to draw a backwards arrow over the word.

Her husband then went on to tell me that she makes her own smoothies to go in her PEG, but despite being told not to, she continues to use nuts in the smoothie, and they get stuck in her tube. So, is this just because she really wants to eat nuts or because she does not understand the consequence of using them and damaging her PEG tube? I think this highlights how valuable a study will be that looks at the eating experience in the home with people who have cognitive decline.

I certainly can see the value of doing this screener for the HCPs working with pwMND – it is so important for HCPs to be aware of how much is being understood with complex and new information such as PEG intervention, diet changes and how to eat via a tube.

It was at this point that my curiosity about cognitive and behavioural changes in MND grew. I wanted to understand how HCPs recognise this symptom, how they assess it clinically and then how they manage it. I also wanted to understand the HCPs’ attitudes to this symptom of MND. Do they feel like it is an important symptom to consider in the greater context of MND?

I also wanted to understand the experiences of families with MND and to learn if they were being educated about this symptom, or indeed if they even wanted to know about it.

From my observations and from investigating the literature, my PhD research changed, from the original plan of conducting a biomechanical investigation of dysphagia in MND, to a qualitative program of research, which aimed to better understand the interaction of cognitive and behavioural changes in MND and dysphagia. I wanted to speak to many more HCPs, from various disciplines and locations, to understand their approaches to assessing and managing cognitive and behavioural changes, as well as their attitudes and comfort levels. Further, I wanted to understand how people living with MND experience care related to both dysphagia and cognitive and behavioural changes, and to learn their preferences for care. I also wanted to understand if cognitive and behavioural changes interacted or impacted on dysphagia management. Specifically, if a pwMND had cognitive and behavioural changes, how did this impact on their ability to manage a swallowing problem in the home, and how did this impact on the carer.

From my preliminary review of the literature in MND, I began to see that there was a gap related to the interaction between cognitive and behavioural changes in MND and dysphagia management. As a speech pathologist, it was important to me to address this potential gap, with an aim of improving care for families with MND, for all the reasons I describe above. To do this, I realised my research studies needed to be exploratory and person-centred to facilitate the gathering of participants' experiences, perceptions, and behaviours. Additionally, a theory driven framework was needed to analyse these data as a way of investigating facilitators and barriers of this phenomenon. Therefore, a qualitative method of enquiry was more aligned to exploring the interaction between cognitive and behavioural changes in MND and dysphagia.

To summarise, the original plan for my program of research was to conduct quantitative biomechanical studies of swallowing in MND. However, given my experiences in the clinic and my learning about cognitive and behavioural changes in MND, the path changed.

> We shall not cease from exploration
> And the end of all our exploring
> Will be to arrive where we started
> And know the place for the first time (Eliot, 1943)

THESIS STRUCTURE

The work completed for this research is presented in the format of a thesis with publications.

There are chapters included in this thesis and the structure is shown in Figure 2:

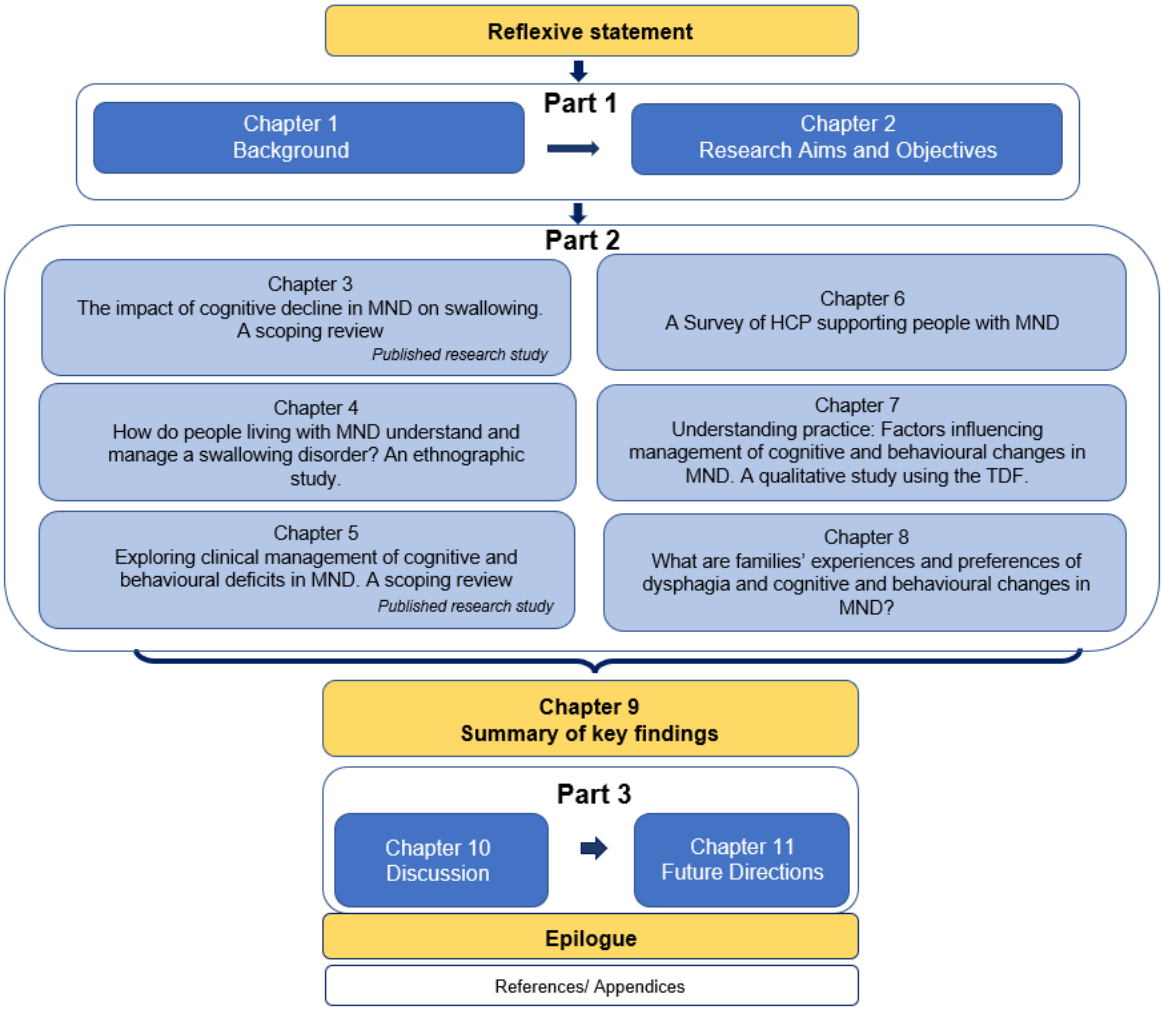

Figure 2: Thesis structure
(MND: Motor Neurone Disease; HCP: Health care professional; TDF: Theoretical Domains Framework). **Image by thesis author.**

PART 1: CHAPTER 1 BACKGROUND

Almost all people with motor neurone disease (pwMND) will develop dysphagia (impaired swallowing) (MND Australia, 2021a; Waito et al., 2017). Dysphagia has negative health consequences in motor neurone disease (MND) related to an increased risk of choking, dehydration, malnutrition and aspiration pneumonia (Miller et al., 2009). Aspiration pneumonia results from food, fluids or secretions entering the lungs via the respiratory tract and is one of the leading causes of death in MND (Oliver, 2019). Consequently, speech pathologists support pwMND by assessing swallowing function and providing management strategies for dysphagia.

A typical management regime for dysphagia may include bolus modifications, where food viscosity, food volume and food textures are adjusted, and/or the thickness of fluids is modified, with the use of thickeners. In addition, behavioural interventions may be suggested. These include strategies such as reducing mouthful volume size, increasing focus and attention during meals, and/or reducing distractions whilst eating. Further, postural modification and manoeuvres may be instructed to support a person to increase the efficiency of their swallowing. When swallowing is too difficult, no longer possible or unsafe via mouth, medical interventions such as enteral feeding procedures may be recommended to sustain life (Rumbach et al., 2016).

All management strategies employed to support a person with dysphagia require cognitive engagement from the pwMND, first to understand instructions and background information in principle and then to enact these in the home environment. Whilst a speech pathologist may make safe swallowing recommendations, as with any management regime or medical intervention, a person has the right to proceed or refuse health care recommendations (Australian Commission on Safety and Quality in Health Care, 2020). Management strategies for dysphagia and the decision-making processes regarding care preferences in

general, require a person to have cognitive capacity to first, understand the problem, then understand their choices related to the problem and then provide informed consent or refusal of recommendations or treatment. However, there are complexities for dysphagia management in MND, as cognitive and behavioural changes are also known to be associated with the disease (Martin et al., 2017).

Approximately 50% of people diagnosed with MND also experience cognitive and/or behavioural changes (Devenney et al., 2015). These changes have been found to occur on a spectrum or continuum with frontotemporal dementia (FTD). Consequently, some pwMND will experience variations of mild to moderate cognitive and behavioural changes, and up to 15% of pwMND will develop symptoms that are consistent with a diagnosis of MND/FTD (Devenney et al., 2015; MND Australia, 2020a).

Cognitive and behavioural changes in MND are important to consider, as they have been documented to impact on a pwMND's insight, concentration, reasoning, problem-solving and planning (Phukan et al., 2007). Further, cognitive and behavioural changes in MND are reported to negatively impact on both a pwMND and their carer's quality of life (Radakovic et al., 2024). Consequently, it is critical to understand if, and how cognitive and behavioural changes in MND interact and impact on dysphagia management.

This thesis aims to extend our understanding of how the interaction between cognitive and behavioral changes in MND impacts on dysphagia management. Additionally, it seeks to explore current management approaches for cognitive and behavioral changes in MND more broadly and to understand the preferences of families affected by MND regarding both dysphagia and cognitive and behavioral changes.

Overview of Chapter 1

In Chapter 1, background information underpinning this program of research will be discussed. Specifically, I will provide a literature review of MND, including the classification of MND types, a brief history of MND, theories related to the aetiology of MND and a description of the complex disease mechanisms. Following this, I will describe diagnostic processes and summarise the symptoms of MND.

Then, typical swallowing function will be described to provide the background information that underpins the section on dysphagia in MND. The dysphagia in MND section will end with a summary of dysphagia management. As dysphagia management strategies require cognitive engagement to enact, I will then provide background information and a description of human cognition. Within this description there will be a focus on executive functioning and social cognition, as these cognitive domains are known to be impacted by MND. Frontotemporal dementia (FTD) will be summarised and the links between FTD and MND will be described. Then a description of cognitive and behavioural changes, known to be associated with MND, will be provided. In addition, an additional non-motor symptom of MND, sensory change, will also be summarised.

The final part of this chapter considers MND management.

Motor Neurone Disease

MND is an umbrella term, for a family of highly heterogenous neurodegenerative diseases that impact on specialised nerve cells or neurones, and their associated axons. MND is a devastating terminal disease which affects approximately 1 per 11,434 Australians (Deloitte Access Economics, 2015). This rate is reported to be relatively high in comparison to other countries. For example, European studies report prevalence rates of 7.9 per 100,000 population (MND Australia, 2021c). However, the global estimate of MND prevalence is unclear, due to wide variability in incidence and prevalence reporting (Chiò et al., 2013; Cronin et al., 2007). It is estimated that the overall global incidence of MND will increase with a predicted ageing population (Chiò et al., 2013; MND Australia, 2021c). Currently, the number of Australians living with MND is estimated to be over 2000 (The Florey, 2024).

The risk of developing MND increases with age, and as such, the lifetime risk is reported to be 1 in 300 by the age of 85 years (Martin et al., 2017). Post symptom onset, approximately 50% of pwMND die within 2.5 years, whereas 20% of pwMND survive between 5 to 10 years after the onset of symptoms. The mean age of death is 69.7 years (The Florey, 2024). Survival rates in MND vary and are dependent on several factors, namely the clinical presentation or phenotype, the rate of disease progression, function of the respiratory system and nutritional status, with higher BMI being linked to longer survival (Dardiotis et al., 2018). The main cause of death in MND is typically related to respiratory failure resulting from respiratory muscle weakness or repeated infections and pneumonia (MND Association, 2022).

The economic cost of MND is substantial, with economic analysis totals showing the cost per person diagnosed with MND to be AUD$1.13 million. Further, as the functional deficits of MND are severe, and the disease is associated with premature mortality and subsequent absence from the workforce of both the pwMND and their carer, the total cost of MND to the Australian community is estimated to be $2.34billion annually (Deloitte Access Economics,

2015). As there is no known cause or cure for MND, continued exploratory and applied research is essential.

Classification of MND

MND involves the progressive loss of upper motor neurones (UMN) (neurones projecting from the primary motor cortex to the brain stem and spinal cord), lower motor neurones (LMN) (projecting from the brainstem and anterior horn of the spinal cord to the muscles), interneurons and glial cells (Philips & Rothstein, 2014). The primary sites of involvement of MND are within the anterior horn of the spinal cord and along the corticospinal and corticobulbar tracts (Arora & Khan, 2020). As a motor neurone's primary function is to innervate muscles, when these motor neurones are damaged or die, their ability to innervate muscles is also impacted. MND may selectively affect only UMNs or only LMNs or involve a combination of both. Consequently, the distinct MND phenotypes are evident through clinical presentation and vary greatly depending on different sites of onset and the rate of progression of neuronal loss (Yoon et al., 2014) as shown in Figure 3.

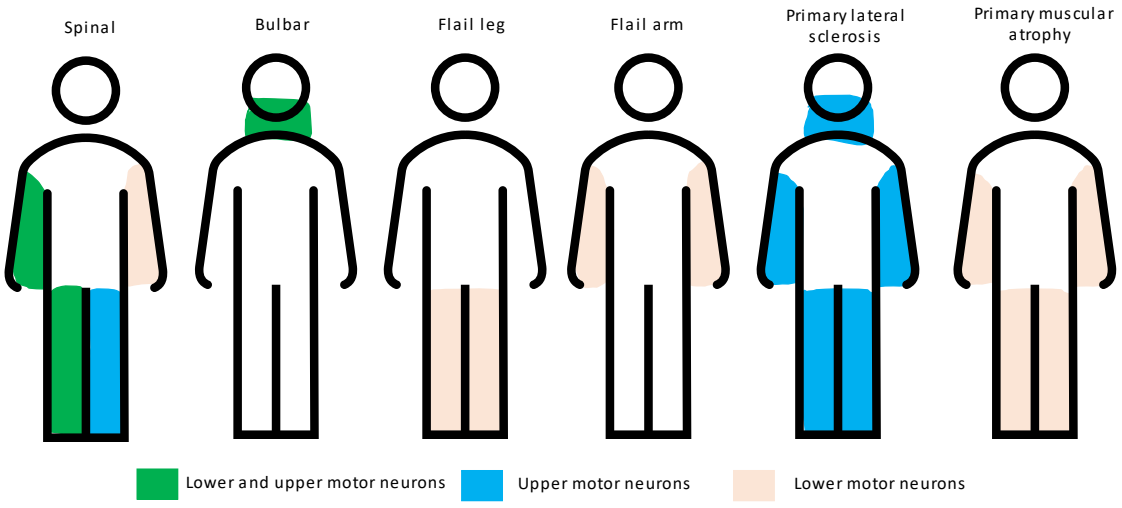

Figure 3: Upper and motor neuron involvement and phenotypes in MND. ***(MND: Motor Neurone Disease).*** **Image by thesis author.**

Clinically, degeneration of UMNs results in the clinical signs of spasticity (increased muscle tone at rest), weakness, hyperreflexia and pseudobulbar affect (Swinnen & Robberecht, 2014), whereas degeneration of LMNs present as flaccidity, paralysis, weakness, hyporeflexia, muscle atrophy and muscle fasciculations (Swinnen & Robberecht, 2014). The variations in clinical presentation contribute to diagnosing a specific type of MND (Talman et al., 2016).

Amyotrophic lateral sclerosis

Amyotrophic lateral sclerosis (ALS) or spinal onset MND, is the most common form of MND, accounting for up to 70% of presentations (MND Association, 2023e; MND Australia, 2024b; Talman et al., 2016), and the average age of symptom onset occurs at 60 years (Swinnen & Robberecht, 2014). The word *amyotrophic* has origins in Greek language, with *a* meaning *"*no*", myo* relating to "muscles" and *trophic* also having a Greek route in the form of *trofí (τροφή)* meaning "nourishment" or "food" (Goodwin, 2003). As such, the literal meaning of *amyotrophic* is "no nourishment to the muscle". Further, *lateral* refers to the anatomical location of the motor neurones within portions of the spinal cord, and *sclerosis* can be defined as "scarring" or "hardening" (RX List, n.d.).

The clinical hallmark of pure ALS, as per the earliest Charcot descriptions (described below) (Swinnen & Robberecht, 2014) involves the presentation of painless focal adult-onset weakness and atrophy, usually in the limb muscles and combining both UMN and LMN signs (Williams, 2013). Subsequent to asymmetrical muscle weakness, spasticity typically develops indicating UMN involvement (Jankovska & Matej, 2021). Rarely, in about 5% of cases, a person will present with initial trunk or respiratory involvement (Kiernan et al., 2011). In addition to the motor signs of ALS, clinically, a person with probable MND may also experience observable fasciculations, describe pseudobulbar affect, weight loss, or cognitive changes. These symptoms are described below within the Symptoms of MND section (Pg. 25).

Bulbar onset MND or progressive bulbar palsy (PBP)

Bulbar onset MND is seen in approximately 20 - 25% of MND cases (Turner et al., 2010; Williams, 2013) and is more common in women (Kiernan et al., 2011; Turner et al., 2010). Bulbar onset MND is associated with a progressive loss of both UMNs and LMNs within the brainstem and corticobulbar tract. Consequently, function of the muscles of the head and neck regions are implicated first. The majority of pwMND who have a bulbar onset MND type report dysarthria as the first symptom, with dysphagia symptoms typically occurring second (Donohue et al., 2023). As the disease progresses, muscle weakness typically spreads to the limbs within 12 months (Williams, 2013), affecting both limb and respiratory function (MND Australia, 2024b). Additionally, changes to secretions and impaired cough function are also common in bulbar onset MND, as well as dyspnoea (shortness of breath), which may be exasperated when a pwMND lays in a supine position. Further, bulbar onset MND is associated with higher rates of cognitive impairment (Portet et al., 2001).

Typically, bulbar onset MND has a faster rate of progression, with pwMND surviving between six months and three years post symptom onset (MND Association, 2023e; MND Australia, 2024b; Wijesekera et al., 2009). Notably, it is reported that most cases of bulbar onset MND will advance to ALS with disease progression (Karam et al., 2010).

Progressive muscular atrophy (PMA)

PMA, (also known as progressive spinal muscle atrophy) is a sporadic, adult onset pure LMN disease (Williams, 2013). It is a rare form of MND accounting for approximately 5% of cases and is more likely to affect males (M: F, 3:1-7:1) (Liewluck & Saperstein, 2015), over the age of 55 years. PMA is a generally considered to be a slower progressing phenotype of MND, with survival times greater than five years (Liewluck & Saperstein, 2015). Consequently, a pwMND, initially diagnosed with an ALS MND, may be re-classified with PMA if there are no UMN clinical signs after four years post symptom onset (Martin et al., 2017).

PMA is associated with neuronal loss in the anterior horn of the spinal cord and consists of the LMN clinical signs of muscle atrophy, muscle weakness, hyporeflexia and fasciculations (Liewluck & Saperstein, 2015). The onset of symptoms in PMA may occur within the upper limbs, lower limbs or axial skeleton (De Carvalho et al., 2007).

Axial onset PMA is associated with a worsened prognosis, as people are more likely to experience earlier respiratory failure (De Carvalho et al., 2007). Predictors of length of survival in PMA are associated with factors that are present at diagnosis, namely the greater number of regions of the body affected, a lower functional vital capacity score or a lower ALS Functional Rating Score (ALS FRS) (De Carvalho et al., 2007). Post-mortem studies have shown that UMN degeneration may be present, however, may remain clinically undetectable (Liewluck & Saperstein, 2015; Rowland, 2010). Additionally, studies have shown that UMN signs may develop within 61 months after PMA diagnosis.

Flail Arm and Flail Leg syndromes

Both Flail Arm Syndrome (FA) and Flail Leg Syndrome (FL) are both atypical forms of LMN MND, which are reported to have subtle or late UMN signs (Menon et al., 2016). Typically, slower progressing, the survival rates in FA and FL are reported to be greater than five years post symptom onset, and prognosis is improved if the progression of the disease does not spread into another region (Kiernan et al., 2011). Currently, there is some debate amongst researchers if FA and FL are variants of ALS MND or distinct disease entities. Both syndromes are frequently misdiagnosed as classic forms of ALS (Kornitzer et al., 2020), and additionally, during early stages of disease, limb onset ALS may be mistaken for a FA or FL syndrome, due to the localisation of weakness (Kornitzer et al., 2020)

First described in 1886 by Vulpian (1886), FA is an atypical presentation of ALS, also known as brachial amyotrophic diplegia (Yang et al., 2015). FA is associated with the LMN signs of symmetrical proximal weakness and atrophy, however, only in the upper limbs (Hübers et al., 2016). In FA, the age of onset is similar to ALS, however, there is a much higher male

dominance with a male to female ratio between 4:1 and 9:1 reported (Hu et al., 1998; Wijesekera et al., 2009). Whilst in FA the functional involvement of lower limbs and the bulbar region are relatively spared (Yang et al., 2015), some report that electrophysiological studies reveal both cortical and peripheral hyperexcitability (Menon et al., 2016). Additionally, at least one UMN sign may occur in up to 50% of people with a FA diagnosis (Yoon et al., 2014).

Discovered by Pierre Marie in 1918 (Zapalska et al., 2023), FL is also referred to as leg amyotrophic diplegia or pseudopolyneuritic variant of ALS (Menon et al., 2016). FL is another rare, atypical presentation of ALS, associated with symmetrical weakness and atrophy to the lower limbs, with absent lower limb tendon reflexes. FL has a much slower progression compared to ALS, with longer mean survival rates reported to be between 6 to 7yrs post symptom onset (Wijesekera et al., 2009). Additionally, UMN signs have been reported to occur in up to 78% of FL cases (Menon et al., 2016). However, different to FA, the male to female ratio in FL is reported in some studies to be 1:1 (Wijesekera et al., 2009).

Primary lateral sclerosis (PLS)

Primary lateral sclerosis is an idiopathic progressive pyramidal form of MND, affecting only UMN (Williams, 2013). PLS accounts for approximately 3% of cases and is a much slower progressing phenotype (Kiernan et al., 2011). This phenotype may be confirmed if there are no LMN signs after four years (Kiernan et al., 2011; Martin et al., 2017). The length of survival with PLS is approximately 10-20 years post symptom onset, and consequently may not be life shortening (Statland et al., 2015). However, as the symptoms of PLS are related to problems with balance, spasticity in the muscles and weakness occurring primarily in the legs, it is associated with significant functional impairment. Additionally, in PLS, urinary urgency or increased frequency may occur (Statland et al., 2015).

There is some debate if PLS and ALS are distinct disorders or occur along an ALS continuum. It is suggested that if people present with spasticity, have an absence of clinical

LMN signs and do not develop muscle wastage within three years from onset, it is more likely to be a pure PLS (Statland et al., 2015).

Whilst the slower progressing forms of MND, such as PLS, are related to longer survival, the increased functional deficits may be associated with higher socioeconomic burden on families living with the disease.

Kennedy's Disease.

Sometimes also described under the umbrella of the MND literature is Kennedy's Disease. This rare inherited disease, named after William R. Kennedy in the 1960s, affects lower motor neurons and results in muscle atrophy and weakness in both the limbs and head and neck (MND Australia, 2024b; Williams, 2013). Kennedy's Disease may also be referred to as spinobulbar muscular atrophy (SBMA) (Spada et al., 1991). The cause of Kennedy's Disease is linked to a genetic mutation, and predominantly affects men. Symptoms of Kennedy's Disease present similarly to MND and thus people may be incorrectly diagnosed. Different to MND, the symptoms associated with Kennedy's Disease can be managed and consequently, this disease is not typically life-limiting (MND Australia, 2024b). However, as with MND, there is currently no cure for Kennedy's Disease.

Despite the description of different types of MND, differential diagnosis of MND types is often difficult due to the co-occurrence of symptoms across MND types. Clinical observations throughout disease progression may result in an initial diagnosis of one type of MND being relabelled to a different type of MND, depending on pathophysiological signs, such as rate of progression, functional deficits and continued survival (Meo et al., 2022).

Earliest observations and a brief history of MND

Although symptoms of limb weakness and muscle atrophy were previously recorded, MND was first referred to as the distinct disease 'Amyotrophic Lateral Sclerosis' by neurologist Jean-Martin Charcot (1825-1893), as per this excerpt from his 1874 lecture series (Duyckaerts et al., 2021; Eisen et al., 2024; Kumar et al., 2011):

> a) progressive atrophy invading the muscles; b) fibrillary contractions which are especially seen in the active period of the atrophy; c) the preservation of faradic contractility that the wasted muscles exhibit to the last moment...Other symptoms are quite foreign to protopathic spinal amyotrophy; first, motor weakness that occurs early and which, if it does not precede atrophy, at least is strikingly evident when the latter is not yet well-developed…The extremities, more or less deprived of their natural movements, are usually in lateral sclerosis affected by rigidity at rest, resulting from what is called continual spasmodic contractures. This phenomenon is absolutely foreign to primary atrophy (Charcot, 1874).

In his work, Charcot pioneered the clinicopathologic approach (or anatomo-clinical method), which describes clinical presentations of living patients with corresponding pathological lesions post-mortem. Involving Charcot's work at the Salpeˆtrie`re Hospital, Paris, France, the descriptive characteristics of MND were recorded and based on clinical observations of twenty patients and five post-mortem studies (Kumar et al., 2011). Through these investigations, Charcot hypothesised that the motor component of the spinal cord consisted of two distinct systems, whereby the subsequent location of a lesion would manifest as a distinct clinical presentation. It was these early studies that contributed greatly to our knowledge of the organisation of the central nervous system. Along with fellow neurologist, Alix Joffroy (1844-1908), Charcot determined that lesions within the lateral column of the spinal cord manifested in contracture and progressive paralysis (now referred to as spasticity, or upper motor neurone signs), whereas lesions in the anterior horn of the spinal cord resulted in muscle atrophy and paralysis (lower motor neurone signs). Through these continued investigations, Charcot also observed that these pathologies frequently co-occurred (Eisen et al., 2024; Goetz, 2000; Rowland, 2001). Whilst further genetic and molecular discoveries related to MND have since been made, Charcot's early clinical and pathological descriptions of MND have largely remained unaltered. Despite this, it was not

until much later in 1941, that wider global awareness of MND occurred (ALS Association, 2024).

In the United States of America, a well-known baseballer for the New York Yankees, Lou Gehrig, received a diagnosis of MND from the Mayo clinic in Rochester, Minnesota in 1939. Gehrig delivered a farewell speech to over 61,000 fans at Yankee Stadium on the 4th of July 1939. He died from the disease on the 2nd of June 1941, at 37 years old (National Baseball Hall of Fame, n.d.). Given Gehrig's high sporting profile, much attention was given to the disease and consequently raised public awareness of MND. As a consequence, for many years after, MND was known as Lou Gehrig's disease, and this term may still be used by some today.

Much later, in 1993, the first gene mutation associated with familial MND was discovered, the superoxide dismutase 1 or 'SOD1' gene (Rosen et al., 1993). Further explanation of this is described in the 'Genetics in MND' section (Pg.15) Importantly, the SOD1 mutation discovery facilitated the engineering of the first MND transgenic SOD1 mouse model. It was this model that was instrumental in furthering our understanding of the MND disease pathogenesis for pharmalogical treatment clinical trials.

After many failed drug trials, in 1995, the drug Riluzole, whilst not a cure, was shown to slow the progression of MND (Lacomblez et al., 2002; Onesti et al., 2017). Riluzole works by blocking the release of glutamate and supressing glutamate activity within cells as well as stabilising sodium channels (Kretschmer et al., 1998; Motor Neurone Disease Association, 2024). Therefore, the pathophysiological target of Riluzole is excitotoxicity. Riluzole was approved for human use by the American Food and Drug Administration board (FDA) and is still the most prescribed medical treatment for MND globally. Riluzole is linked to extended survival, with estimates of prolonged life being an average of approximately three months.

In 2015, a second pharmalogical treatment, Edaravone, initially approved for the treatment of strokes, underwent clinical trials for application in MND treatment. As Edaravone is an antioxidant and is thought to be a free radical scavenger, its pathophysiological target is to reduce oxidative stress, and when used in conjunction with Riluzole, was initially shown to slow functional decline in pwMND (Rothstein, 2017). Edaravone was first approved for use within Japan and Korea, and in 2016 was also approved by the FDA. Additionally, around this time, Edaravone was approved for use in Canada, Switzerland, China, Indonesia, Thailand and Malaysia. However, it was not until February 2023 that the drug was approved for use in Australia by the Therapeutic Goods Administration (TGA) under the product name RADICAVA®. Edaravone is also not a cure for MND, and in addition, there have been mixed findings as to its efficacy (Witzel et al., 2022). However, some studies have reported Edaravone to be effective at slowing MND progression in the earlier stages of the disease (Jiang et al., 2022).

The most promising advancement in the treatment of MND came in April 2023, when the FDA expiated approval of the drug Tofersen for the treatment of MND. Tofersen was developed to treat SOD1 variant MND. An antisense oligonucleotide, Tofersen specifically targets mutated RNA (Figure 4) produced from the mutated SOD1 gene, to prevent toxic protein aggregation from occurring. Consequently, the neuropathophysiological target of Tofersen is genetic mutation and the subsequent development of aberrant proteins. The administration method for Tofersen is via lumbar puncture.

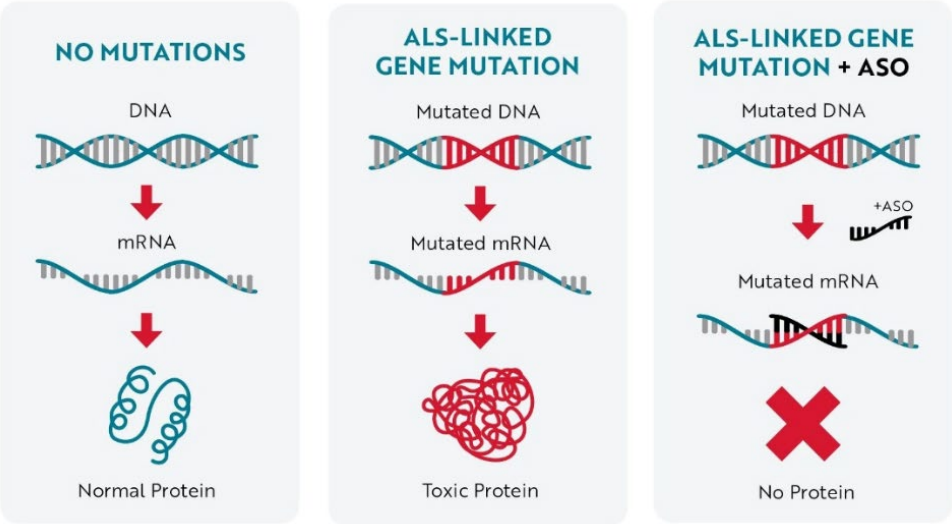

Figure 4: How does Tofersen work?
Figure retrieved from: https://www.als.org/navigating-als/living-with-als/fda-approved-drugs/tofersen. Reproduced with permission from the ALS Association.

Clinical trials of Tofersen are ongoing and aim to evaluate the effectiveness of this drug to slow MND progression. Additionally, investigations are exploring if Tofersen can prevent or slow symptoms of disease in SOD1 carriers who are pre-symptomatic. However, at the time of writing this thesis, Tofersen was not approved for use within Australia. Here, I highlight some additional references for specific reading about the efficacy of Tofersen and the ongoing clinical trials (Benatar et al., 2022; Miller et al., 2020; Miller et al., 2022).

Aetiology

MND is highly heterogeneous, and as such, the aetiology of MND remains unknown (MND Association, 2023e; M. D. Wang et al., 2017). It is reported that the majority (up to 90%) of MND cases are sporadic in origin, making identification of a distinct cause difficult. However, there are some factors that are known to be associated with an increased risk of developing MND. These risks include age and being male (M. D. Wang et al., 2017). Specifically, males over the age of 65 years are overrepresented in MND (Martin, 2017). Whilst some genes have been linked to familial MND, these only account for approximately 10% of MND cases (Acevedo-Arozena et al., 2011; Kiernan et al., 2011).

Genetics in MND

Many diseases show complex disease patterns, ranging from familial disease with autosomal dominant inheritance pattern to simplex families, where only one person develops the disease, leading to assumptions of sporadic disease origin (Al-Chalabi & Lewis, 2011). In MND, there are numerous genes now known to be associated with developing the disease, some with clearer familial links, accounting for approximately 5 to 10% of cases (Al-Chalabi et al., 2014). Additionally, it is suggested that approximately 20% of MND cases may have affected relatives, identified through pedigree studies, or more extensive population-based investigations (Al-Chalabi et al., 2014), which, due to lower gene penetrance, may be assumed to be of sporadic onset. However, it is important to acknowledge that the gene mutations found to be associated with MND are not always easily linked to familial origins. Gene testing in people who have no family history of MND, reveal approximately 20% have a genetic test result that requires a clinical response (Al-Chalabi et al., 2014). Consequently, there are a high proportion of seemingly sporadic onset cases, with no known direct familial links (Brown et al., 2021).

There are approximately 50 known gene mutations that have been linked to MND, however four genes account for 60% of familial cases including C9orf72, SOD1, TARDBP, and FUS (Dubowsky et al., 2023). For the purpose of this thesis, the two most commonly linked gene mutations, the SOD1, accounting for approximately 10-30% of familial cases (Berdyński et al., 2022; Dubowsky et al., 2023), and the C9orf72 gene accounting for approximately 40% of cases (Dubowsky et al., 2023) (Figure 5) will be described.

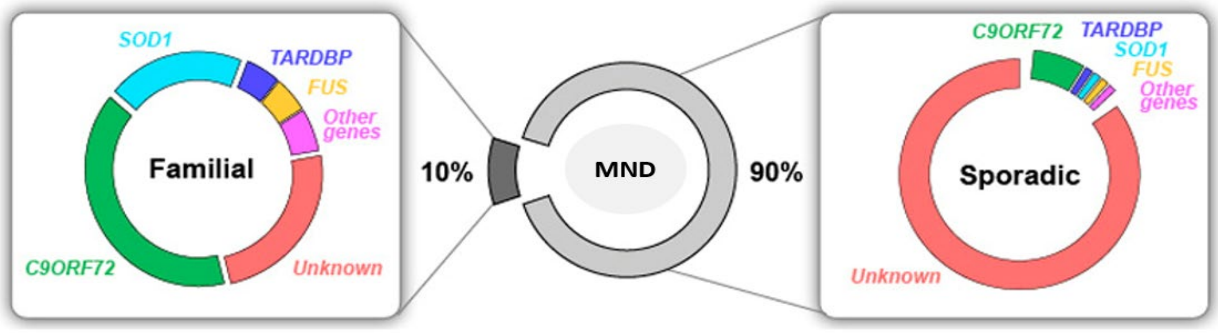

Figure 5: Sporadic and familial onset MND showing genes known to be associated with MND. Image retrieved from Laferriere and Polymenidou (2015). Reproduced with permission under Attribution-Non Commercial-Share Alike 4.0 Internation. ***(MND: Motor Neurone Disease)***

The first gene mutation linked to MND was discovered in 1993, when a mutation in the superoxide dismutase 1 gene, or the SOD1 gene, was found (MND Australia, 2020b; Rosen et al., 1993). SOD1 is a widely expressed protein, and as such, mutations in SOD1 can destabilise these proteins creating cellular degeneration. There are over 180 known disease-causing variants to the SOD1 gene, and in MND this gene mutation is considered high penetrance (Al-Chalabi et al., 2014) meaning there is high probability that someone carrying this mutated gene will develop clinical signs of disease (Zlotogora, 2003). Notably, there have been 170 mutations of the SOD1 gene identified to cause MND (Laferriere & Polymenidou, 2015). Through investigations using mouse models, it was discovered that the SOD1 gene can selectively affect motor neurones with deficits in tongue and limb function recorded (Acevedo-Arozena et al., 2011). In familial onset MND, the SOD1 gene has been linked to between 10% to 30% of cases within families (Dubowsky et al., 2023).

A further variant of the SOD1 gene is the SOD1 A5V variant, which results in a more rapid and aggressive form of MND disease progression (Opie-Martin et al., 2022). The average age of onset for the SOD1 A5V is 49 years, and the average length of survival is less than two years post symptom onset. Another variant, the SOD1 I114T mutation, is associated with slower progressing cell degeneration and a slower rate of disease progression. This

variant is also linked with later disease onset, with 50% of people experiencing symptom onset around 60 years of age.

The C9orf72 gene mutation was discovered in 2011 (De Marchi et al., 2023). The pathological process of C9orf72 involves a hexanucleotide repeat expansion G_4C_2 (e.g., GGGGCC). The function of the non-pathological C9orf72 gene is unclear but is suspected to play a role in autophagy and endosomal trafficking, as well as the regulation of endoplasmic reticulum stress (Zhang et al., 2014). Whilst this repeat gene expansion occurs within all humans, in the case of both MND and FTD, the repeat sequence is expanded hundreds and even thousands of times (Gijselinck et al., 2018). It is currently not clear how these repeat expansions contribute to both MND and FTD (Zhang et al., 2014) disease. However, these expansion sequences will result in wide and varied clinical presentations, depending on the length of the repeat expansion. The C9orf72 gene mutation is associated with the most common cause of familial MND and is also the gene mutation that is most linked to the MND-Frontotemporal dementia (MND-FTD) spectrum (Devenney et al., 2015).

To summarise, familial genetic cases account for approximately 10% of MND cases. The remaining 90% of MND cases are seemingly sporadic in origin, however pedigree studies may reveal familial links. It is hypothesised that the onset of MND symptoms may involve a multi-step disease process, involving six steps (Al-Chalabi et al., 2014).

Multi-step disease model.

Multi-step models of disease onset were originally developed in the cancer epidemiology context and more recently, these models have been used to investigate the aetiology of MND (Al-Chalabi et al., 2014). Within this research, it is hypothesised that six distinct genetic and/or environmental exposures are required for MND to occur, and it is the sixth exposure which will result in triggering the onset of MND (Al-Chalabi et al., 2014). This hypothesis is formed on the basis of several disease anomalies. Specifically, MND is an adult-onset disease. However, many people, known to have the genetic mutation, which is present from

birth, will live a healthy life with disease onset occurring between the ages of 50 to 70 years. Consequently, they experience variations in location of symptom onset and of disease progression. Currently, there has been no genetic or environmental factors found to be associated with the site of symptom onset (Al-Chalabi et al., 2014). Paradoxically, others, also born with the mutated gene, have been found to remain healthy into old age, never developing MND (Al-Chalabi et al., 2014). It is unknown why pathological genetics, present from birth are expressed later in adult life in some, and not at all in others. Consequently, whilst this six-step model may provide a better understanding of MND aetiology, the identification of these steps is exceptionally difficult, with an infinite number of combining factors. Therefore, beginning with investigations that consider the known genetic mutations may help in narrowing down and identifying additional disease factors. For further reading on this multi-step model in MND, please refer to the following studies (Al-Chalabi et al., 2014; Vucic et al., 2020).

Environmental risk factors

There are several theories of environmental risk factors related to the sporadic onset of MND. Environmental risk factors in MND are difficult to identify, given that there are an almost infinite number of environmental factors and combinations of these factors to investigate. To further add to this complexity, designing large scale studies to investigate all possible environmental factors are cost prohibitive. Additionally, environmental investigations rely on history taking and a person's accurate account of information, which is not always a reliable source. As a result, only a select number of assumed environmental risk factors have been investigated.

Exposure to heavy metals, typically linked to an occupation, have been shown to increase the risk of developing MND (Martin et al., 2017; MND Australia, 2021c; M. D. Wang et al., 2017). The most commonly reported metal associated with MND is lead (Kamel et al., 2005), followed by mercury, although mercury has not been extensively studied (M. D. Wang et al.,

2017). Other heavy metals that are linked as an environmental risk of developing MND include selenium, magnesium aluminium, manganese and cadmium (M.D. Wang et al., 2017). In addition to heavy metal exposure, chemical exposure, specifically the chemical pesticides of pentachlorobenzene and cischlordane are reported to have some association with an increased risk of developing MND (M. D. Wang et al., 2017).

The numerous chemicals found in cigarette smoke have been linked to neurotoxic effects (Alonso et al., 2010). Further, cigarette smoke has been found to contain lead, which has previously been linked to an increased risk of MND, as described above. In addition, formaldehyde (Weisskopf et al., 2009) is present in cigarette smoke and has also previously been associated with an increased risk of MND. Overall, however, the links between cigarette smoking and MND have been inconclusive and do not support a strong association. However, it is suggested that smoking may pose as a higher risk factor for women (Alonso et al., 2010)

Additional theories about the cause of MND have been linked to physical activity, traumatic brain injury, military service and exposure to cyanotoxins such as α-Amino-β-methylaminopropionic Acid (BMAA) associated with blue green algae (Thomas, 2024; M. D. Wang et al., 2017). Further, caffeine exposure and interestingly, even chicken soup have been investigated as a potential protective mechanism in MND. However, within investigations of environmental factors there is variable evidence and difficulties replicating studies to demonstrate causality (Martin et al., 2017; MND Association, 2023d).

Retrovirus in MND

Studies have shown that 59% of pwMND have a retrovirus signature in blood testing, compared with only 5-8% of the general population (Douville et al., 2011; Li et al., 2015). The theories of retrovirus in MND involve inheritance, not infection, and an active virus can trigger MND onset. It is reported that the human endogenous retrovirus type K (HERV-K) is likely the cause, as this retrovirus has been shown to be toxic to motor neurones, specifically

in mice models (Li et al., 2015).

Current clinical trials are investigating the ability of pharmalogical interventions to switch off retroviruses. Specifically, Triumeq, a drug that has been successful in the treatment of human immunodeficiency virus (HIV) infection, is being investigated for repurposed use in MND (Gold et al., 2019). Phase 2a trials, completed in Australia, have shown Triumeq (administered via a single once-daily tablet) to be well tolerated and safe for pwMND, and there are positive signs that Triumeq may suppress genetic reactivation. Consequently, Phase 3 Randomised Double-Blind Placebo controlled trials are underway, with 27 locations currently recruiting in the United Kingdom, Ireland, Netherlands, Slovenia, Spain, Sweden, New Zealand and Australia. The reader is referred to (Dubowsky et al., 2023; Gold et al., 2019; Macquarie University Australia, 2024) for further information on Triumeq.

Complex pathological mechanisms of MND

Similar to cancer, MND is associated with profound clinical, prognostic, neuropathological and genetic heterogeneity. Consequently, as our understanding of MND mechanisms evolve, the concept of MND as being a single type of disease is increasing challenged (Turner et al., 2013). Many microscopic changes that reflect multifaceted and complex pathophysiological processes have been identified as associated with MND, however, currently it is not clear which mechanisms are the most important to prioritise through targeted treatment methods. The processes known to be involved in MND include oxidative stress, generation of free radicals, neuroinflammation, excitotoxicity, mitochondrial dysfunction, impaired protein homeostasis, impaired DNA repair, dysregulated nucleocytoplasmic transport, aberrant RNA metabolism, dysregulated vesicle transport, axonopathy and glial cell dysfunction (Kiernan et al., 2011). Figure 6 depicts these key factors. It was the discovery of mutations within the TAR DNA Binding Protein 43kDa (TARDBP, encoding TDP-43), observed to be mislocalised, and consequently pathological in up to 97% of MND cases, which has provided critical insights into MND (Martin et al., 2017).

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Review at https://doi.org/10.1016/S0140-6736(10)61156-7

Figure 6: Mechanisms of disease linked to MND are multifactorial and complex and operate through inter-related molecular and genetic pathways.
Figure adopted from Kiernan et al. (2011)
(MND: Motor Neurone Disease)

TDP-43

TDP-43 is an essential, ubiquitous human protein expressed in many tissues. It is encoded by the TARDBP gene (Jo et al., 2020) and plays an important role in normal cellular function (Heyburn & Moussa, 2017). In healthy cells, TDP-43 is localised within the nucleus of neuronal cells but has the ability to shuttle between the nucleus and cytoplasm, to carry out a range of cellular functions (Jo et al., 2020). For example, M-RNA transport, M RNA splicing and MRNA stability (Jo et al., 2020). Therefore, cells are sensitive to alterations in TDP-43 levels (Heyburn & Moussa, 2017).

Positive ubiquitinated cytoplasmic inclusions of TDP-43 have been found to occur in approximately 90% of MND cases (Jo et al., 2020). Specifically, the protein has been shown to unfold and aggregate within the cytoplasm, becoming sticky. It is suggested that this

protein aggregation interrupts the essential functioning of the other cellular proteins and consequently results in neuronal cell death (Martin et al., 2017). Further, clinical observations of pwMND have shown that increases in TDP-43 pathology, in combination with motor neuron cell death, contribute to disease progression (Jo et al., 2020; Suk & Rousseaux, 2020). Developing further understanding of TDP-43 pathology in MND is an active area of ongoing research.

To summarise, MND is an umbrella term for a series of highly complex, adult onset, terminal neurodegenerative diseases. Once symptoms begin, neurodegeneration is often rapidly progressing and consequently, the functional impact of MND on people is devastating. To add to the complexity, multiple genetic mutations and the identification of multiple disease mechanisms make understanding MND more difficult. Consequently, diagnosing MND is also highly complex.

Diagnosis

Currently, there is no singular test that can diagnose MND (Williams, 2013) and as such diagnosis is based on clinical signs. As the onset of MND symptoms is often insidious, delays with a referral to neurology services have been described (O'Brien et al., 2011a). Instead, people may initially be referred to other specialities, such as physiotherapists to treat reported limb weakness, an ENT or speech pathologist for changes to speech for bulbar onset signs (O'Brien et al., 2011a). On average, diagnosis of MND is reported to take on average 12 months between symptom onset to a definite diagnosis (MND Australia, 2021c).

If MND is suspected, however, best practice suggests that an immediate referral is necessary to a specialised neurologist to undertake several investigations to support diagnosis, and offer further support (Williams, 2013). As such, a detailed clinical history and examination is vital as part of the diagnostic process of MND, to both discover and differentiate between upper motor neuron and lower motor neuron signs through observation

(Yoon et al., 2014). A further part of the diagnostic process is a series of tests, such as urine tests, electromyography (EMG), nerve conduction studies (NCS), functional magnetic resonance imaging (fMRI) of the brain and spinal cord, blood tests, lumbar punctures and muscle biopsy. These tests, along with the assessment of clinical features, are important as they allow for other diseases to be ruled out. In addition to the above, regular and ongoing monitoring is needed, as MND can only be diagnosed if motor symptoms progress (Brooks, 1994). To add to the diagnostic complexity, health care professionals (HCPs) may be hesitant to diagnose MND until they are certain, for fear of missing other treatable conditions (Williams, 2013). Consequently, a formal diagnosis of MND is often delayed. Despite the diagnostic complexity, misdiagnosis rates of MND are relatively low, reported to be between 7-8% of cases (Davenport et al., 1996; Traynor et al., 2000).

The criteria for diagnosing MND as per the El Escorial Criteria (Brooks, 1994) consists of the following:

- A presence of both UMN and LMN clinical signs AND
- progressive spread of symptoms within one region or spread to one of the other four regions of the body (Bulbar, Cervical, Thoracic, Lumbosacral) WITH
- an absence of electrophysiological evidence applicable to other disease mechanisms AND
- an absence of neuroimaging evidence of other disease processes which may account for the observed clinical signs.

Biomarkers

Biological markers, or biomarkers, are an objective and observable sign that can be used to detect or monitor the progression of disease (Rogers, 2022). Biomarkers are important in disease treatment as they support development of effective therapies (Rogers, 2022). Currently, however, there are no biomarkers that can definitively diagnose MND, although

neurofilaments (NFs) are emerging as a promising biomarker for the clinical assessment of many neurodegenerative diseases, including MND.

Neurofilaments are a cytoskeletal protein found within neurons, that function in abundance, to contribute to the internal scaffolding and structure of the neuron. When neuronal axons are damaged or die, NFs are released into biological fluids in proportionate levels to the degree of axonal damage (Sun et al., 2020). Consequently, elevated levels of NFs will be found in many neurodegenerative diseases via both blood (plasma) and cerebral spinal fluid (CSF). Whilst NFs have been shown to be highly specific to neuroaxonal damage, they have not, as yet, been found to be specific to disease (Gordon, 2020).

Testing of NFs was previously only possible by cerebral spinal fluid sampling, obtained via invasive lumbar puncture. However, with the advent of highly specialised and sensitive automated technologies, it is now possible to assess and quantify proteins via plasma taken with a simple blood test (Benatar et al., 2020).

A sub-unit of NFs, neurofilament light chains (NfL), have been reported to be significantly higher in pwMND when compared to healthy controls (Sun et al., 2020). Additionally, NfL levels have been positively correlated with rate of disease progression. For example, higher serum NfL levels were reported in MND compared to those with a diagnosis of PMA and PLS type MND (Benatar et al., 2020). Subsequently, it was reported that increased levels of NfL in blood predict ALS type MND severity and progression (Sun et al., 2020)

Whilst the precise role of serum NFs as a biomarker in MND is not yet fully defined, ongoing investigations are considering the potential use NFs in clinical practice to track disease progression via re-testing methods. Additionally, serum NFs biomarkers hold promise for use within clinical trials as a possible pharmacodynamic biomarker, to determine which experimental therapeutics progress from Phase II to Phase III clinical trials (Benatar et al., 2020; Zucchi et al., 2020)

For further reading about the utility of serum NfL as a biomarker in MND, please refer to (McCluskey et al., 2023)

An additional fluid biomarker, which is being investigated as a potential diagnostic biomarker in MND, has shown some promise in detecting the presence of proteins containing cryptic exon-excluded neoepitopes in CSF indicating TDP-43 dysregulation in MND FTD patients (Irwin et al., 2024). Specifically cryptic hepatoma-derived growth factor-like protein 2 (HDGFL2) has been shown to accumulate in increased levels in MND FTD. This is an important finding, as this biomarker is also showing promise of detection during the pre-symptomatic phase of known C9orf72 gene mutations. For further information the reader is directed to (Irwin et al., 2024)

Symptoms of MND

Symptom onset in MND is insidious (Kiernan et al., 2011). The early symptoms of MND are typically mild, and consequently may be initially ignored, and are often mistaken for other conditions (NHS Inform, 2024; O'Brien et al., 2011). The most common clinical presentation of MND is the loss of motor function, characterised as weakness. Collectively, loss of motor function impacts on a multitude of other functions including mobility, speech, respiration and chewing and swallowing. Additionally, symptoms of insomnia, fatigue, cramps, pain and depression and anxiety are also reported by pwMND.

Mobility

In limb onset MND, the first symptoms typically occur in either the arms or legs as focal weakness or stiffness (MND Association, 2023e). Initially, this weakness or stiffness may be described by a person as reduced grip strength, increasingly dropping objects, impaired ability to raise the arms or shoulders, an increase in trips or falls and/or an observation of reduced size of the affected limb (muscle atrophy) (MND Association, 2023e). The initial experiences of weakness and stiffness are usually painless in the early stages of disease

onset (MND Association, 2023e; NHS Inform, 2024). Additionally, fasciculations in the muscles may co-occur with the above symptoms. Importantly though, fasciculations in isolation are not reported to occur in MND (Youn & Scelsa, 2020). All of these abovementioned symptoms will worsen as MND progresses.

Respiration

As all types of MND progress, weakness to the respiratory muscles will eventuate (Heffernan et al., 2006). Both expiratory and inspiratory muscles are affected, which involve the intercostal muscles of the ribs and the diaphragm muscle. In the initial stages of respiratory function, dyspnoea (breathlessness or shortness of breath) occurs. As MND progresses, dyspnoea worsens, and the discomfort of this increases with physical activity, or when lying in a supine position, impacting on sleep function. With disease progression, respiratory insufficiency results in impaired gas exchange, and consequently can cause interrupted sleep, morning headaches, decreased appetite, impaired concentration or confusion, irritability and fatigue (MND Association, 2023e). Additionally, a pwMND will have a weakened ability to cough, increasing the risk of aspiration and respiratory tract infections (Heffernan et al., 2006). As such, respiratory function is a significant clinical indicator of survival (Heffernan et al., 2006). Further, in rarer cases (approximately 5%) (Kiernan et al., 2011), respiratory onset MND may occur and a pwMND may experience dyspnoea as the initial symptom of the disease. Towards the end of life, severe respiratory insufficiency occurs, and pwMND may experience increased levels of anxiety associated with respiratory difficulty. Towards the end stages of life pwMND may have difficulty remaining awake to due to depleted oxygen levels. As such, the majority of deaths in MND are caused by eventual respiratory failure (Heffernan et al., 2006). Often, a pwMND will sleep deeply prior to passing away, and death in MND is typically described as being peaceful.

Pseudobulbar affect

Pseudobulbar affect (PBA), also referred to as emotional lability, is a common symptom in MND with up to as many as 50% of pwMND experiencing this symptom (MND Australia, 2020a). PBA is generally associated with bulbar onset, spinal onset and PLS types of MND (Floeter et al., 2014), given their UMN and corticobulbar tract involvement (Finegan et al., 2019; MND Australia, 2020a). PBA is described as pathological laughing or crying, that is inappropriate, exaggerated or unrelated to a person's current emotional state (Finegan et al., 2019). This symptom causes considerable psychosocial impacts for both a pwMND and their families, and is associated with increased social isolation, and reduced quality of life (Andersen et al., 2012).

PBA is linked with a pathological disruption to the neuronal networks involved in the regulation and inhibition of emotional output (Finegan et al., 2019). Specifically, in MND, this is suggested to be the corticopontocerebellar tracts (Floeter et al., 2014) and involves the fronto-cortical regions, pons and associated cerebellar connections (Finegan et al., 2019). However, there is considerable clinical variation in PBA presentation across pwMND, and therefore, PBA in MND is said to occur on a spectrum (Miller et al., 2011). Within affected individuals, the symptoms of PBA, however, remain fairly consistent.

Weight loss and malnutrition in MND

Weight loss is also common in MND and is linked to a faster rate of disease progression and has a negative effect on survival (Dharmadasa et al., 2017; MND Australia, 2021b; Ngo et al., 2019). Additionally, premorbid lower body mass index (BMI) levels are linked to a higher risk of developing MND and also worse prognosis (Ngo et al., 2019). The causes for weight loss in MND are multifactorial. First, weight loss may be related to impaired swallowing function (further explained in Dysphagia in MND on Pg.43) or feeding difficulties due to

impaired limb function. Second, pwMND have been shown to experience hypermetabolism (Steyn et al., 2018).

Hypermetabolism is defined by a significant increase in resting energy expenditure (Vaisman et al., 2009) and is reported to occur between 25% and 70% of pwMND. This percentage is reported to increase to 100% of people with a familial type of MND (Steyn et al., 2018). Increases in resting energy expenditure may be related to cooccurring impairments in respiratory function (Vaisman et al., 2009) as with disease progression, greater energy expenditure is required to support breathing. Third, weight loss in MND is also likely to occur as a consequence to changes in appetite (Holm et al., 2013).

Changes in appetite in MND are not completely understood. However, appetite is known to decrease as the disease progresses, and results in a poorer prognosis (Holm et al., 2013; Ngo et al., 2019). Additionally changes to appetite in MND have been reported to be associated with a decrease in respiratory function (Holm et al., 2013). Appetite changes in MND may also be associated with psychosocial factors, such as depression or low mood. Additionally, it is reported that changes to both metabolic rates and eating behaviours in MND may be associated with the FTD disease continuum (Ahmed, Irish, et al., 2016). Overall, more work is needed to understand both appetite and weight loss in MND (MND Australia, 2021b).

Speech

Speech is a highly coordinated motor task, relying on muscular control of the whole speech mechanism (Tomik & Guiloff, 2010). As MND impacts on the innervation of these muscles, dysarthria (impaired speech) is common, occurring in approximately 80% of cases, and occurring earlier in bulbar onset MND (Tomik & Guiloff, 2010). Dysarthria in MND may be further categorised as flaccid or spastic dysarthria, depending on the underlying pathology. Flaccid dysarthria results from LMN pathologies and is associated with the muscles involved

in speech wasting and becoming weaker. Therefore, a pwMND may present with slowed movement of the tongue, lips and pharyngeal muscles, impacting on speech (Makkonen et al., 2018). In contrast, with an UMN spastic dysarthria, slowness of movement and weakness will be observed without wasting (Makkonen et al., 2018). Additionally, as MND can be associated with both UMN and LMN pathology, a mixed flaccid-spastic type of dysarthria can also occur.

Dysarthria in MND impacts on a pwMND's intelligibility, and speech is typically characterised as slow and laborious, with monotone pitch, imprecise articulation and increased hypernasality (Hanson, 2011; Tomik & Guiloff, 2010). Given MND is also associated with impaired respiratory function, a pwMND may also present with reduced volume if there is insufficient expiration to support phonation (Duffy, 2019; Makkonen et al., 2018). Furthermore, innervation of the arytenoid and lateral cricoarytenoid muscles (CNX) may be impacted, and consequently, vocal fold adduction may be impacted (Duffy, 2019). This will result in changes to vocal quality, as the vocal folds are not able to approximate or vibrate efficiently. As such, a pwMND may present with a breathy or strained and strangled voice quality (Hanson, 2011). Additionally, disruptions to prosody (rate and rhythm of speech) may be observed (Hanson, 2011).

Dysarthria typically worsens as MND progresses. In some, dysarthria will cause a person's speech to become unintelligible or progress to anarthria (absence of intelligible speech), and consequently, communication will need to be supported with alternative approaches, such as low and/or high-tech augmentative and alternative communication devices (AAC) (Hanson, 2011). AAC aims to support or replace spoken communication. However, implementation of AAC is highly dependent on the pwMND's mobility and cognitive ability and consequently, require specialist speech pathology assessment and regular review (Mackenzie et al., 2016).

Salivation

It is common for pwMND to experience altered salivation, with sialorrhoea (excessive thin saliva), xerostomia (dry mouth) or thick tenacious saliva described. This is more likely to occur in the initial stages of bulbar onset MND and may occur with disease progression in spinal onset MND. Salivation is explained further in the Dysphagia in MND section (Pg.43)

Secondary impairments

Pain associated with MND is reported to be nociceptive pain, rather than neuropathic pain, and may be as a consequence of reduced mobility, rather than directly caused by MND (Lau et al., 2018). With disease progression and continued neuronal degeneration, increased spasticity, muscle atrophy and paralysis of the affected limbs will occur. Subsequently, a pwMND may experience aching or stiffness in the joints, with increased levels of pain or discomfort. Joint pain is thought to be associated with non-uniform disease progression, indicating there is an imbalance in function of the agonist-antagonist muscle groups, which results in increased joint stress, and subsequent pain (Burke et al., 2023). For example, adhesive capsulitis, more commonly known as ‘frozen shoulder’, is described (Shanahan et al., 2020). PwMND often also experience debilitating pain and reduced range of shoulder joint movement related to weakness of the periscapular muscle (Burke et al., 2023). Frozen shoulder is reported in approximately 20% of pwMND, making this a common secondary impairment of MND (MND Association, 2023e). Treatment typically involves a suprascapular nerve block (Shanahan et al., 2020).

An additional secondary impairment associated with MND is constipation, which may also be as a consequence of reduced mobility (MND Association, 2023e; Yamamoto et al., 2024). However, medications, dehydration or enteral feeding methods (Yamamoto et al., 2024) are also described as contributing to constipation in MND.

Swallowing

Lastly, almost all pwMND will develop dysphagia (impaired swallowing) over disease progression (MND Association, 2023b; MND Australia, 2021a). To better understand the symptom of dysphagia, as it relates to MND, first typical swallowing function will be described.

Swallowing

Like breathing, swallowing is a physiological act vital to survival. On average, humans swallow between 500 and 700 times per day and like breathing, we continue to swallow whilst sleeping (Speech Pathology Australia, 2024). Swallowing is necessary for nutrition and hydration and additionally, serves to clear secretions (saliva) from the oral cavity (Panara et al., 2023). As well as facilitating delivery of essential nutrition and hydration, swallowing also contributes greatly to quality of life.

Human relationships and interactions are often accompanied by shared eating and drinking experiences. Shared mealtimes are associated with meaningful opportunities for families to bond, build connections and enhance communication, and have been shown to be a positive contributor to mental health outcomes (Heikkilä et al., 2022; Middleton et al., 2022; Smith et al., 2020). In addition, celebrations such as weddings, religious ceremonies and holiday family gatherings typically involve friends, family and loved ones coming together to bond over a shared meal. Whilst most humans associate enjoyment with the act of eating and drinking, many have never considered the physiological act of swallowing.

Swallowing is a highly complex sensorimotor process that requires the integration and coordination of sensory input from the peripheral nervous system and motor output within the central nervous system (Steele & Miller, 2010). In healthy swallowing, the action is swift, highly coordinated, and precisely timed. As such, swallowing relies on a large neuronal network, with continuous sensory input to modulate motor output. Each swallowing event engages over 26 pairs of striated muscles and nine of the 12 cranial nerves (Matsuo &

Palmer, 2013; Seikel et al., 2018). Adding further complexity to swallowing physiology, the oral and pharyngeal phases of swallowing share the anatomical structures of the head and neck with parts of the respiratory tract (Malone & Arya, 2020). Consequently, the act of swallowing is associated with two vital functions: i. to transport food, fluid and oral secretions via the digestive tract into the stomach for digestion (Ertekin & Aydogdu, 2003; Malone & Arya, 2020) and ii. to protect the airway from ingested materials (Ertekin & Aydogdu, 2003; Malone & Arya, 2020). The protective function of swallowing will be discussed in more detail within the dysphagia in MND section (Pg. 43) to follow.

The neurophysiology of swallowing is multifaceted, highly complex and as yet not fully understood. With the advancement of imaging technology, specifically fMRI (Glover, 2011), our understanding of regional brain activation, event timing and changes to brain metabolism has drastically increased. Task activation fMRI studies have allowed further understanding of the neurophysiology of swallowing. Where previously thought to be a reflex, studies have collectively shown that swallowing is mediated through involvement of both cortical and subcortical regions as shown in Figure 7. During a swallowing event, fMRI imaging shows bilateral activation within the primary motor cortex, somatosensory cortex, internal capsule, basal ganglia, cerebellum and brain stem (Ertekin & Aydogdu, 2003). As such, pathology in these areas can result in impaired swallowing function (Malandraki et al., 2011).

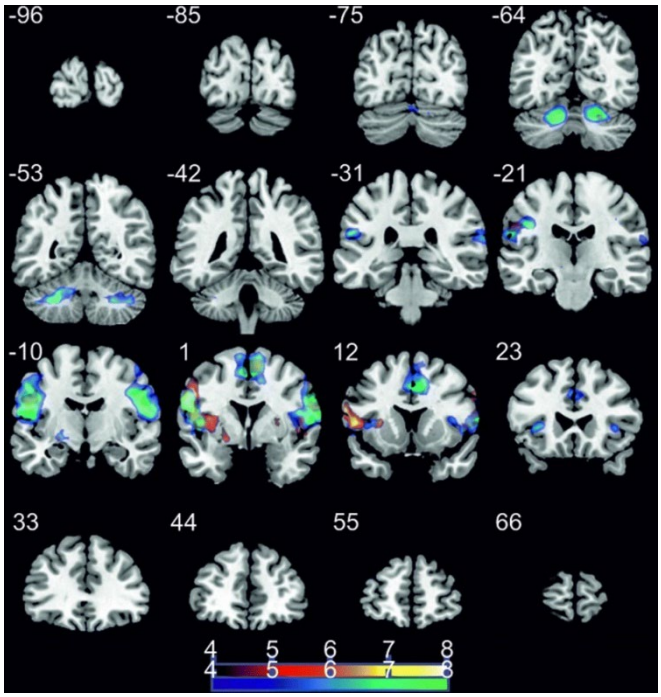

Figure 7: Cortical and cerebellum activation during swallowing as shown on fMRI imaging. Image retrieved from Kober et al. (2019). Reproduced with permission Creative Commons CC BY license. ***(fMRI: Functional magnetic resonance imaging)***

Pairs of cranial nerves involved in swallowing include the sensory-only nerves of CNI (Olfactory), CNII (Optic) and CNVIII (Vestibulocochlear), the mixed cranial nerves: CNV (Trigeminal), CNVII (Facial), CNIX (glossopharyngeal) and CNX (Vagus), as well as the motor-only nerves, CNXI (Accessory) and CNXII (Hypoglossal). CNI and CNII originate from the cerebrum, whereas the remaining cranial nerves involved with swallowing emerge from the brainstem, with CNV and CNVII emerging from the pons, CNVIII emerging from the pontomedullary junction and CNXI and CNXII emerging from the medulla (Malone & Arya, 2020). The role of each cranial nerve will be described in more detail within the phases of swallowing sections (Pg. 36). In the following section, the role of the swallowing central pattern generators (CPGs) will be described, as their intricate function in sensory-motor integration can be affected by MND.

Located within the brainstem and spinal cord, CPGs consist of a dense network of neurons and interneurons. These neuronal networks are solely responsible for enacting stereotypical sequences of motor commands that are associated with rhythmic movements, such as those

expressed in breathing, walking, standing, and chewing (Bucher et al., 2015). Additionally, CPGs are associated with the less rhythmic movements that occur in mastication, sneezing and vomiting (Koga & Fukuda, 1997; Lang, 2009). Swallowing is a also highly complex, patterned movement.

Swallowing is one of the most complicated motor patterns humans enact, due to significant bilateral motor function across the body's midline, coordinating both the activation and inhibition of approximately 26 pairs of striated muscles (Dodds, 1989). As shown in Figure 8, the swallowing CPGs, involving several brainstem motor nuclei (V, VII, IX, X, XII), are located within two main areas of the medulla oblongata of the brainstem, specifically, the dorsal medulla within the nucleus tractus solitarius (NTS), and the ventrolateral medulla, just anterior to the nucleus ambiguous (NA) (Jean, 2001).

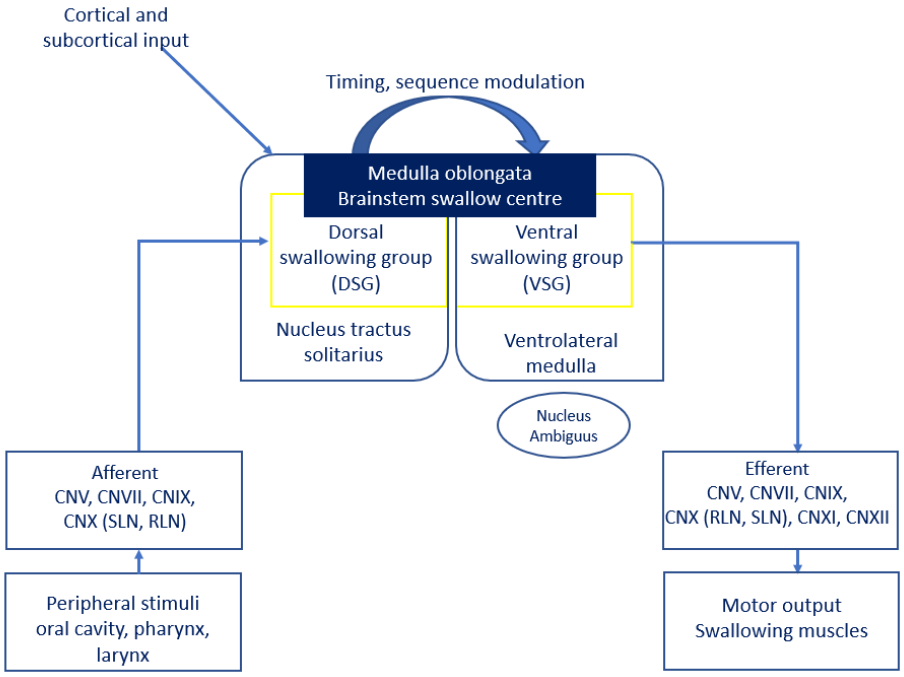

Figure 8: Schematic diagram of swallowing central pattern generator located in the brainstem. *(DSG: Dorsal swallowing group; VSG: Ventral swallowing group; CNV: Trigeminal nerve; CNVII: Facial nerve; CNIX: Glossopharyngeal nerve; CNX: Vagus nerve; CNXI: Accessory nerve; CNXII: Hypoglossal nerve; RLN: Recurrent laryngeal nerve; SLN: Superior laryngeal nerve).* Image created by thesis author and adapted from Langton-Frost (2024)

Within the dorsal medulla and adjacent reticular formation, the dorsal swallow group (DSG) neurons are located. The DSG contains the generator neurons which function to trigger, shape and time the sequential motor pattern of both the oropharyngeal and oesophageal phases of swallowing (Jean, 2001). Further, the ventral swallowing group of neurons (VSG) function to distribute the motor plan out to various motor neurone pools involved in swallowing (Jean, 2001). A descending motor pattern from the cortex may initiate a volitional swallow, but the sequence of movement, and subsequent pattern of muscle activation, is not part of this descending motor information. Instead, this is thought to be controlled at the level of the brainstem within the swallowing CPGs (Langerman et al., 2018; Langton-Frost et al., 2024).

The motor output involved in swallowing is altered in response to sensory input from peripheral areas during mastication and the act of swallowing itself (Langton-Frost et al., 2024). Sensory input from the oral cavity, pharynx and larynx is relayed to the swallowing CPGs in the brainstem, specifically the NTS of the medulla oblongata (Jean, 2001). Information such as texture or viscosity, volume, temperature and taste all contribute to bolus-specific modifications of the swallowing motor output from the DSG via the VSG and relevant cranial nerves (Langerman et al., 2018; Langton-Frost et al., 2024). Therefore, any impairment to the intricate network of brainstem neuronal activity can result in impaired swallowing function.

Whilst the act of swallowing is a highly complex and an interdependent mechanism, conceptually, swallowing can be described in phases that relate to the position of the bolus (ingested material) as it moves from the oral cavity towards the stomach.

Anticipatory Phase (Pre-Oral phase)

The first phase of swallowing is referred to as the pre-oral or anticipatory phase (Matsuo & Palmer, 2009). This phase of swallowing may not be consistently reported in the literature as the first phase of swallowing, as it does not involve the movement of food or fluid from the oral cavity into the stomach (Shune et al., 2016). However, this is an important phase, as it is a preparatory phase, where sensory input related to smell, sound, anticipated taste, volume, viscosity and temperature is processed (Matsuo & Palmer, 2009; Rubesin, 2021; Shune et al., 2016). Pre-oral motor, cognitive and psychosocial factors of this anticipatory phase all influence the oral and pharyngeal phases and are, consequently, important to consider in oropharyngeal swallowing assessments.

Anticipatory sensory information not only contributes to a person's preparedness to eat (Shune et al., 2016) but also to the modulation of motor output required to execute an efficient swallow (Matsuo & Palmer, 2009). Each of the five special senses, olfaction, vision, tactile perception, hearing and gustation, as well as the sixth sense of proprioception, or the body's ability to sense movement, action and location in relation to itself and its environment, are important factors. In addition, cognition (e.g., awareness and motivation) contributes to a person's ability to recognise food and their willingness to eat (Shune et al., 2016). During the pre-oral phase, there is integration of auditory, olfactory and visual stimuli, which stimulates autonomic responses, increasing salivation and gastric peristalsis/secretions, as well as upregulation of the motor nuclei involved in swallowing in response to sensory information, in preparedness for the oral phase. Any impairment in this phase can lead to uncoordinated swallowing (Patey et al., 2012)

Oral Preparatory Phase

The oral preparatory phase of swallowing involves the structures and processes within the oral cavity and commences when food or fluid reaches the oral (buccal) cavity. Under volitional control (Jean, 2001), the oral phase is highly variable and depends on factors such as hunger, levels of fatigue, environment, and consciousness.

The anterior border of the oral cavity consists of the orbicularis oris muscle (lips) (CNVII), which provides a seal and prevents anterior spillage of oral contents. The Trigeminal nerve (CNV) innervates both the jaw opening and jaw closing muscles, which facilitates bolus admittance and bolus containment during mastication. Laterally, the cheeks, which consist of the buccinator muscles (CNVII) contribute to oral control of a bolus by contracting to press against the teeth. The superior border of the oral cavity consists of the palatine process of the maxilla and the palatine bones, which contribute to the anterior hard palate. The posterior soft palate is muscular and consists of the palatoglossus, palatopharyngeus, levator veli palatini muscles, musculus uvulae (CNX), and the tensor veli palatini (CNV). The inferior border of the oral cavity, or the floor of mouth, consists of four muscles: the anterior belly of the digastric (CNVII), geniohyoid (CNV), stylohyoid (CNV) and mylohyoid (CNV) (Toth & Lappin, 2019; von Arx et al., 2017). The floor of mouth provides stability for the tongue during mastication (chewing).

The tongue is a muscular organ (Sasegbon & Hamdy, 2017), which is instrumental in deglutition and speech (Seikel et al., 2018). The tongue body consists of four paired intrinsic muscles, the superior and inferior longitudinal, verticalis and transverse which all function to change the shape of the tongue (CNXII) (Seikel et al., 2018). The extrinsic muscles of the tongue, the hyoglossus, styloglossus, hyoglossus (CNXII), and the palatoglossus (CNX) (Malone & Arya, 2020), function to change the position of the tongue within the oral cavity. For sensation, the anterior two thirds of the tongue relays general sensory information such as temperature, pressure and pain (CNV). Taste receptors are largely concentrated in fungiform papillae on the anterior two thirds of the tongue and the circumvallate papillae in the sulcus terminalis (AlJulaih & Lasrado, 2019). The special sense of taste to the anterior two thirds of the tongue is relayed via the chorda tympani (CNVII) (Seikel et al., 2018). In addition, retronasal olfaction contributes to taste sensation, as during mastication, air, containing odour producing molecules, is forced via the nasal cavity to stimulate olfactory receptors (CNI) (Blankenship et al., 2019). The posterior third of the tongue is innervated by

CNIX for both general sensation and the special sense of taste (Dotiwala & Samra, 2018). Collectively, it is the intricate movements of the tongue, which function to move the bolus around the oral cavity (Matsuo & Palmer, 2013) during bolus preparation.

Mechanical processing within the oral cavity is aided by chemical processing to prepare food to a consistency that is primed for swallowing and digestion. During the oral preparatory phase, the tongue functions to position food to make contact with the teeth. Mastication involves rhythmic movements of the mandible (jaw), facilitated by the temporomandibular joint and surrounding muscles that receive innervation from CNV (Matsuo & Palmer, 2013). Further, the buccinator muscles contract to prevent food spilling into the lateral sulcus (between the cheek and the teeth), and the orbicularis muscle contributes to creating an anterior seal of the oral cavity (Logemann, 1984). To aid in both mastication and taste, saliva, consisting of the enzyme amylase, is produced by the sublingual, submandibular and parotid glands, and has a dual function. First, saliva moistens and mixes with the bolus, assisting the tongue in bolus preparation (Alhajj & Babos, 2023; Matsuo & Palmer, 2013). Second, saliva acts as a solvent and supports transport of taste substances to taste bud receptors within the tongue (Matsuo, 2000). During the oral preparatory phase, the airway is open, allowing for nasal breathing, and both the larynx and pharynx are in a resting position.

Chemoreceptors and mechanoreceptors within the oral cavity detect and provide sensory feedback on bolus characteristics to enable a modulated motor response during mastication, including information regarding the readiness for bolus propulsion by the tongue. The final process within the oral phase relates to bolus transport.

Oral transport phase

The initiation of swallowing begins with the tongue (Logemann, 1984). The bolus is gathered on the dorsal portion of the tongue, and the tongue shape changes to close off the anterior portion of the oral cavity and open the posterior portion, to allow for bolus passage into the oropharynx (Sasegbon & Hamdy, 2017). The anterior tongue approximates with the hard

and soft palate (Rubesin, 2021), squeezing the bolus posteriorly toward the faucial arches (Logemann, 1984). Concurrently, the posterior portion of the tongue flattens, to create a slope for the bolus to move along (Rubesin, 2021). The tongue then propels the bolus into the oropharynx with force (Dodds et al., 1990). The bolus entering the oropharynx coincides with the pharyngeal wall moving anteriorly.

Pharyngeal Phase

The pharyngeal phase of swallowing is described as a transport phase, as the bolus travels through the muscular pharyngeal chamber (Sasegbon & Hamdy, 2017). In addition to bolus transport, the pharyngeal phase functions to ensure the airway is protected during swallowing (Lang, 2009). Hence during this phase of swallowing, respiration temporarily ceases (Sasegbon & Hamdy, 2017).

The pharynx is a vertical chamber which spans from the base of the skull to the upper oesophageal sphincter (UOS) (Donner et al., 1985). As shown in Figure 9, the pharynx can be described in three parts, the superior nasopharynx, which spans from the base of skull to the soft palate, the oropharynx which spans from the soft palate to the pharyngoepiglottic fold. The most inferior portion of the pharynx is referred to as both the hypopharynx or laryngopharynx and spans from pharyngoepiglottic fold to the UOS (Donner et al., 1985).

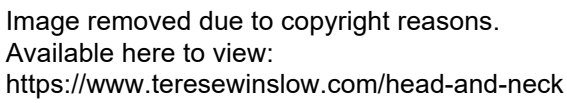

Figure 9. Anatomy of the pharynx showing the superior nasopharynx, medial oropharynx and inferior hypopharynx regions. Image retrieved from https://www.teresewinslow.com/head-and-neck.

The pharyngeal phase of swallowing is predominantly involuntary and consists of a series of pre-programed coordinated and fast events (Shaw & Martino, 2013), as the tail of the bolus reaches the faucial arches. Food properties, such as taste, texture, temperature and viscosity are sensory mechanisms which contribute to bolus-specific modifications (Dodds et al., 1990) of the swallowing motor output and modulate the pharyngeal response. A swallow response is initiated from various points, including the posterior portion of the tongue, epiglottis, aryepiglottic folds, pyriform sinuses, palatine tonsils, soft palate and posterior pharyngeal wall (Dodds et al., 1990). Reconfiguration of both the laryngeal and pharyngeal structures occurs during swallowing, which functions to both protect the airway and propel the bolus through the pharynx, towards the upper oesophageal sphincter (UOS). Once a swallow commences, the motor sequence is irreversible, and there is an overlapping

sequence of both activation and inhibition of muscles of the palate, pharynx, larynx and oesophagus (Jean, 2001). Further, the soft palate contracts (tensor veli palatini, CNV) and elevates (levator veli palatini, CNX), which creates a seal between the oropharynx and nasopharynx (Sasegbon & Hamdy, 2017). This process is referred to as velopharyngeal closure and functions to prevent nasal regurgitation (Perry, 2011).

Following velopharyngeal closure and tongue base retraction, the hyolaryngeal complex initiates superior and anterior movement, commonly referred to as hyolaryngeal excursion. Additionally, the intrinsic muscles of the larynx, the lateral, oblique, and transverse cricoarytenoid muscles, innervated by the recurrent laryngeal nerve (CNX) adduct the vocal folds, closing the glottic space and protecting the airway. The larynx and hyoid bone move superiorly and anteriorly, which functions to move the laryngeal inlet out of the path of the descending bolus, and to pull open the tonically contracted UOS (Sasegbon & Hamdy, 2017). Hyolaryngeal excursion also serves to support epiglottic deflection.

The epiglottis is a cartilage of the larynx, which inverts to complete the closure of the laryngeal vestibule and diverts the bolus towards the pyriform sinus (Rubesin, 2021). Pharyngeal shorteners function to proximate the UES closer to the bolus head. This shortening of the pharyngeal space reduces pharyngeal volume and increases intrapharyngeal pressure. Combined, these movements all contribute to airway protection and the bolus being admitted across the now open UOS into the proximal oesophagus (Malone & Arya, 2020). As the bolus descends through the pharynx, the superior, middle and inferior pharyngeal constrictor muscles contract, producing a contractile wave of muscle contraction which functions to exert pressure on the bolus tail to clear the pharynx of bolus residue (Malone & Arya, 2020).

Oesophageal Phase

The final phase of swallowing is entirely involuntary and is referred to as the oesophageal phase. This phase of swallowing typically begins with a strong recontraction of the UOS muscles and the initiation of oesophageal peristalsis (Miller & Britton, 2011), with rings of smooth muscle (CNX and the sympathetic trunk of both the cervical and thoracic region) (Seikel et al., 2018) contracting in a wave like sequence to propel the bolus (Patel & Thavamani, 2023). Specifically, distension of the oesophageal lumen by the bolus within the oesophagus activates sensory neurons. Consequently, contraction of smooth muscle occurs upstream of the bolus tail and relaxation of smooth muscle occurs from the head of the bolus. This sequence of contraction and relaxation permits bolus propulsion along the length of the oesophagus and admittance through the lower oesophageal sphincter, into the stomach for digestion (Dodds, 1989).

Any breakdown of any of the described parts of the swallowing process is known as dysphagia (Malone & Arya, 2020). Therefore, dysphagia is a symptom of a variety of underlying pathologies, but not in itself a primary medical condition (Etges et al., 2014).

Dysphagia in MND

The word dysphagia has its origins in the Greek language with the prefix *dys* meaning "impaired" or "disordered" and *phaegin* meaning "to eat" (Online Etymology Dictionary, 2017). More specifically, dysphagia can be classified as oral dysphagia, oropharyngeal dysphagia or oesophageal dysphagia, to denote the specific location or phase of functional breakdown.

The aetiology of dysphagia is wide and varied. However, a risk factor for dysphagia is related to neurodegenerative disease, due to the interruption of neural activity that is involved in the swallowing process. For example, it is suggested that almost all pwMND will develop dysphagia as the disease progresses (Romero-Gangonells et al., 2021).

Dysphagia in MND is neurologic in origin, associated with the progressive degeneration of both the excitatory and inhibitory swallowing neurons that control the CPG, causing disruption to the innervation of the muscles involved in swallowing. As is a common theme when describing the presentation of MND, the presentation of dysphagia in MND is also highly variable. There are, however, some consistent characteristics of swallowing changes that are linked to the disease and can be described within the following phases.

Oral Phase difficulties in MND

The most commonly reported difficulties within the oral phase of swallowing in MND are related to impaired bolus preparation and impaired bolus propulsion (Waito et al., 2017).

Bolus preparation

Innervation of the orbicularis oris (lip muscles) (CNVII), may be impaired, resulting in reduced muscle tone and muscle atrophy. This, consequently, results in an inefficient anterior seal of the oral cavity, preventing a pwMND from efficiently containing bolus or liquid within the oral cavity. Consequently, food, fluid and oral secretions spill from the mouth anteriorly. Furthermore, impaired mastication contributes to inadequate bolus preparation.

Impaired mastication in MND is multifactorial. Specifically, decreased or absent innervation to the jaw opener and jaw closer muscles (CNV) may prevent adequate range of movement of the mandible (lower jaw), which results in insufficient bit force. Further, innervation to the lateral and medial pterygoid muscles (CNV), which facilitate a rotary chewing motion to break up and grind food, may also be interrupted (Romero-Gangonells et al., 2021; Waito et al., 2017). Further, impaired innervation to the complex contribution of both intrinsic and extrinsic tongue muscles, which function to both change the shape and move the tongue (CNXII), within the oral cavity (Epps et al., 2020) results in reduced coordination, reduced strength, muscle atrophy and reduced range of motion. Combined these factors all contribute to inadequate bolus preparation. Further, it is common for people with bulbar symptoms to experience altered salivation, with sialorrhoea (excessive thin saliva), xerostomia (dry mouth) or thick tenacious saliva described (James et al., 2022; MND Association, 2023c). Decreased autonomic salivation via the parotid, sublingual and submandibular glands may also contribute to impaired chemical mastication of foods, inhibiting adequate bolus preparation (Peyron et al., 2018). When a pwMND experiences impaired swallowing, sialorrhoea and impaired lip seal, this frequently results in drooling (James et al., 2022; Pearson et al., 2021). Drooling has been shown to have negative health outcomes, linked with an increased risk of aspiration pneumonia, and skin irritations around the mouth. Additionally, drooling is associated with negative psychosocial outcomes (James et al., 2022) with increased distress and embarrassment described by pwMND (Pearson et al., 2021).

Impaired palatal innervation and impaired tongue control to contain the bolus during the oral phase may result in oral contents prematurely spilling into the pharynx. As the glottic space is open during the oral phase, the risk of pre-swallow aspiration is increased. Additionally, impaired innervation of the palatal muscles, specifically, the levator palatini and the tensor palatini (CNV and CNX respectively), may prevent adequate closure of the nasopharynx.

Bolus propulsion

During the subsequent pharyngeal phase of swallowing, a bolus must be propelled by the tongue towards the pharynx. Bolus propulsion relies on adequate strength and range of motion of the tongue. In MND, the intrinsic and extrinsic muscles of the tongue may be weak, spastic and/or atrophied (Romero-Gangonells et al., 2021). Therefore, the tongue may not have adequate range of movement or force to propel the bolus backwards toward the oropharynx. Additionally, if the tongue is not able to adequately contain and propel the bolus, oral residue in the oral cavity is common (Waito et al., 2017).

Pharyngeal Phase difficulties in MND

In MND, the autonomic transportation phase of swallowing is described as mistimed and uncoordinated. Specifically, the swallowing trigger is often delayed or absent (Ertekin et al., 2000), and the opening and closing of the cricopharyngeal muscle of the UOS is often mistimed to the location of the bolus (Briani et al., 1998; Ertekin et al., 2000a). For example, the UOS has been shown to have shorter opening duration or prematurely close whilst the larynx is still in an elevated position (Ertekin et al., 2000). Additionally, atypical bursts of motor activity of the cricopharyngeal muscle have been observed via EMG, occurring during hyolaryngeal elevation (Ertekin et al., 2000). This impaired functioning of the cricopharyngeal muscle is associated with impaired bolus admission through the UOS. Further, reduced pharyngeal constrictor muscle contraction during swallowing results in inadequate pressure generation and leads to reduced bolus clearance through the pharynx. Consequently, bolus residue may remain in the pharynx after the swallowing action is complete. Notably, pharyngeal residue creates an increased risk of post-swallow aspiration, as at the completion of a swallowing, the laryngeal vestibule returns to its resting state and breathing resumes. As such, pharyngeal residue may spill into the now unprotected airway. Consequently, impaired hyolaryngeal elevation and excursion and insufficient glottal closure increase aspiration risk in MND both during and after swallowing (Waito et al., 2017).

Sequential swallowing studies in MND (as opposed to single discrete swallows) have shown arhythmic and irregular pharyngeal swallowing patterns (Aydogdu et al., 2011), suggestive of brainstem CPG dysfunction (Aydogdu et al., 2011). Specifically, CPG neuronal degeneration within the NTS results in impaired triggering, timing and modulation of the sequential swallowing response (Aydogdu et al., 2011).

Aspiration

Dysphagia in MND increases the risk of aspiration. Aspiration may occur in both healthy and unwell people (Kollmeier & Keenaghan, 2023) and involves food, fluids, oral secretions or gastric contents enter the larynx, passing through the vocal folds, and into the lungs. However, the consequences of aspirating are highly variable and dependant on the frequency, type and amount of material aspirated (Ficke et al., 2020), as well as a person's functional status and comorbidities (Hayashi et al., 2014).

Dysphagia increases the risk of developing the bacterial infection known as aspiration pneumonia (Hayashi et al., 2014). In MND, dysphagia often cooccurs with respiratory impairment, substantially increasing the risk of developing aspiration pneumonia. However, this generally occurs later in disease progression (Sorenson et al., 2007). This risk of developing aspiration pneumonia in MND is further complicated, as preventative measures generally recommended, such as being active, are reduced in MND due to impaired mobility. This further increases a pwMND's risk of developing respiratory complications (Ficke et al., 2020). Dysphagia in MND is critical to manage, as aspiration pneumonia in MND is associated with shortened survival (Sorenson et al., 2007) and impaired respiratory function and chest infection are the usual cause of death in MND (MND Association, 2021a)

Further, silent aspiration is the term used when aspiration occurs with no clinical evidence of discomfort, distress or an accompanying cough response (Ramsey et al., 2005), and this has been found to occur in MND (Briani et al., 1998). This suggests a potential disruption to upper airway sensory receptors which contribute to a reflexive cough motor response.

However, further studies are needed that investigate sensation in MND (Robison et al., 2022)

Psychosocial consequences of dysphagia in MND

In addition to the health implications that are associated with dysphagia, there are also psychosocial consequences (Lisiecka et al., 2021; Tabor et al., 2016). PwMND, who also have dysphagia, describe reduced eating pleasure and restricted participation in social situations. For example, drooling, repeated coughing when eating in public and anterior spillage of food have associated with feelings of embarrassment (Motor Neurone Disease Association of NSW, 2018). Furthermore, reduced enjoyment of eating is described both in relation to being fearful of choking and modified diet restrictions limiting food choice (Tabor et al., 2016).

Choking is rarely a cause of death associated with MND, however there is increased risk. As such, pwMND are fearful that choking may occur due to impaired respiratory function that can impact on the efficiency of a reflexive cough (Lisiecka et al., 2021; MND Association, 2024) and impaired pharyngeal function as outlined above. Further, when a pwMND feels increased anxiety, stress or fatigue, this can further decrease swallowing function (MND Association, 2024)

Additionally, it is reported in the broader dysphagia literature, that dysphagia is associated with increased care-giver burden (Namasivayam-MacDonald & Shune, 2018). Carers report increased thought regarding meal preparation, social isolation, negative impacts to their daily routine and worry about adequate nutritional intake for their loved one. This caregiver burden is reported to increase relative to the functional deficits of the person they care for. For example, carers report higher levels of burden when providing care for a person whose main intake is via enteral feeding processes. Specifically, the burden was described as increased feelings of fear and anxiety related to increased responsibility (Namasivayam-MacDonald & Shune, 2018).

Assessment methods

Assessment of swallowing function in MND aims to determine the presence or absence of disordered swallowing, as well as learning about the preferences of a pwMND to manage functional changes to their eating and drinking. Through a comprehensive information gathering process, an appropriate and individualised treatment plan can be devised.

Contemporary assessment for dysphagia includes a combination of a clinical swallowing examination (CSE), and instrumental swallowing assessments, consisting of a videofluoroscopic swallow study (VFSS) (Etges et al., 2014), a flexible endoscopic evaluation of swallowing (FEES) (Etges et al., 2014) and high-resolution pharyngeal manometry.

CSE for dysphagia is a holistic, yet subjective swallowing assessment (Etges et al., 2014), which consists of a combination of client and carer report of the swallowing difficulty, medical history taking, an oromotor examination and oral trials to assess swallowing function. An oromotor examination is conducted to assess cranial nerve function and identify pathological signs, with a focus on CNV, CNVII, CNIX, CNX and CNXII. The clinical signs of muscle atrophy, muscle weakness, and fasciculations indicate LMN involvement, and weakness with spasticity is indicative of UMN involvement (McDermott & Shaw, 2008a). Additionally, the head and neck structures are observed and palpated with the aim of identifying asymmetries, dentition, secretions, oral hygiene and oral mucosa. Further, as part of a CSE, oral trials of food and fluids are conducted, if appropriate. These oral trials aim to identify clinical signs of aspiration and to determine optimal food and fluid consistencies to best support safe and efficient swallowing. Additionally, there are validated dysphagia assessment tools such as the EAT-10 (Plowman et al., 2016) and SwalQoL questionnaires, which are dysphagia symptom specific outcome instruments, that enable a subjective score to be allocated by a person's self-report of their swallowing function (Belafsky et al., 2008; Romero-Gangonells et al., 2021)

The findings from these combined assessments may prompt further objective instrumental assessment, such as a FEES, VFSS or high-resolution manometry. The aim of these additional investigations is to determine if aspiration is occurring, identify premature spillage into the pharynx, and identify post swallow pharyngeal residue or bolus flow obstruction through the UOS.

In summary, dysphagia in MND is complex and is implicated in all stages of swallowing. Dysphagia may result in reduced swallowing efficiency and reduced swallowing safety (Waito et al., 2017) and increases the risk of malnutrition, dehydration, and aspiration (Ruoppolo et al., 2013). In addition, dysphagia negatively affects quality of life (Vesey, 2017) and increases caregiver burden (Aoun et al., 2013; Geng et al., 2017; Ng, Talman, & Khan, 2011). The main cause of death in MND is related to respiratory failure (Govaarts et al., 2016; Vesey, 2017), and dysphagia compromises an already at-risk respiratory system, contributing to respiratory infections. In addition, pwMND report experiencing anxiety and fear related to choking (Oliver & Turner, 2010). Therefore, pro-active and well-informed dysphagia management is critical.

Management and interventions for dysphagia in MND

The primary goal of dysphagia management is to support a person to receive adequate nutrition and hydration, and to maintain quality of life related to enjoying foods (Ueha et al., 2023; Vose et al., 2014). All management plans are individualised and based on a comprehensive swallowing assessment, the pwMND's current swallowing function, and their individual preferences (Ueha et al., 2023). Treatment plans are devised to improve the efficiency and safety of swallowing to enable a pwMND to continue to eat via mouth for as long as possible.

Dysphagia management in MND consists of both rehabilitative and compensatory strategies. Compensatory strategies aim to improve immediate swallowing efficiency and are further sub-categorised as postural strategies, diet modification and bolus control techniques.

Postural strategies aim to alter the swallowing biomechanics and maximise movement of the bolus through the oral and pharyngeal cavities. These strategies involve postural adaptations that are dependent on the underlying pathophysiology. These adaptations include chin tuck, head rotation, head tilt or neck extension (Ueha et al., 2023). Additionally, sitting in an upright position when eating and using a pillow to support posture during meals maybe suggested. However, postural strategies are highly dependent on a pwMND's mobility and as such, need to be regularly reviewed and adjusted as MND progresses. Importantly, implementation of postural strategies also requires a sufficient level of cognitive function to first understand, and then to implement consistently whilst eating and drinking.

In addition to postural adaptations, dietary modifications may be recommended to support safe swallowing, and are a common management strategy to manage dysphagia in MND (MND Association, 2023b). The International Dysphagia Diet Standardisation Initiative (IDDSI) is an evidence-based dietary modification framework, which consists of a continuum of 9 levels, and describes standardised food texture and fluid viscosity (Cichero et al., 2017) (Figure 10).

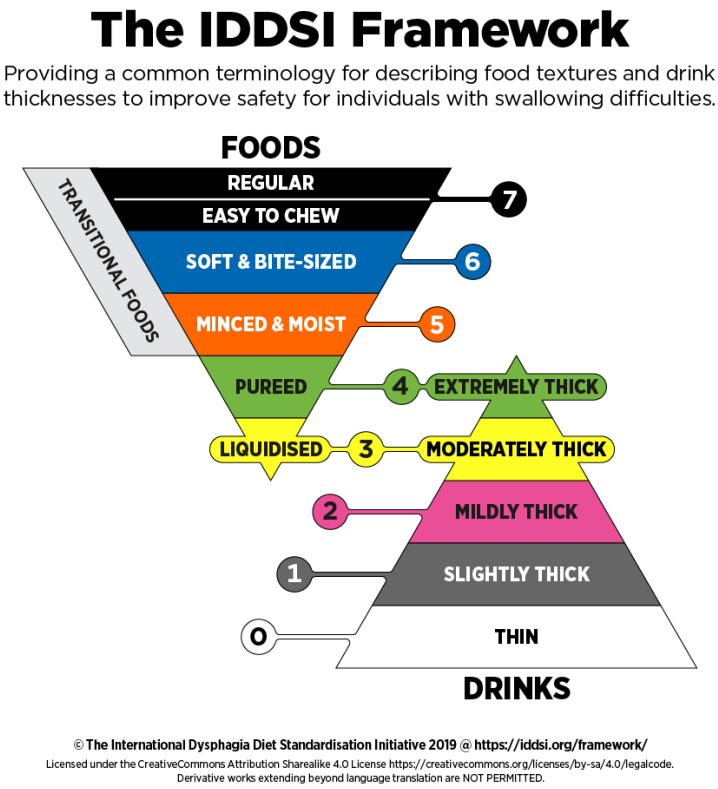

Figure 10: The International Dysphagia Diet Standardisation Initiative (IDDSI). 2019. Dysphagia management food texture modification framework. © The International Dysphagia Diet Standardisation Initiative 2019 @ https://iddsi.org/framework. Image reproduced with permission. Licensed under the CreativeCommons Attribution Sharealike 4.0 License https://creativecommons.org/licenses/by-sa/4.0/legalcode. Derivative works extending beyond language translation are NOT PERMITTED

Additionally, bolus control techniques involve controlling both the rate of intake and size of bolus (Ueha et al., 2023). Further, whilst a pwMND has adequate tongue control, a lingual sweep (moving the tongue around the oral cavity) may be recommended to clear oral residue post swallow. Additionally, if a pwMND has reduced tongue function, yet remaining upper limb control, a finger sweep (placing a finger in the mouth and moving around the oral cavity) post swallow may be suggested. In combination with the above, environmental changes may also be suggested, such as concentrating when eating and avoiding distractions such as eating in front of a television or eating in noisy environments.

In MND management a speech pathologist may operate within a MDT. When considering dysphagia management specifically, a speech pathologist will potentially consult with a gastroenterologist, dietician, occupational therapist and nursing staff (Lau et al., 2018). Collectively these HCPs support a pwMND to eat and drink safely, whilst ensuring sufficient caloric intake, preventing weight loss, and managing the timing of medical interventions, such as enteral feeding procedures, if eating is no longer comfortable or possible via mouth. In this context, dieticians support a pwMND to ensure appropriate caloric intake (Lau et al., 2018), occupational therapists provide support with modified eating and drinking utensils (Lau et al., 2018), and gastroenterologists provide information on and perform PEG procedures. Optimally, discussions with pwMND and family members regarding decision-making for enteral feeding, namely a percutaneous enteral gastrostomy (PEG) procedure are facilitated via joint MDT meetings.

Enteral Feeding

When a pwMND has severe dysphagia, severe fatigue or impaired limb function impacting on eating (McDermott & Shaw, 2008), enteral feeding is typically suggested (Labra et al., 2020). Enteral feeding is used in MND management to provide nutritional support, aiming to prevent malnutrition, dehydration and weight loss, as well as providing an administration method for medications. A gastrostomy procedure, such as percutaneous endoscopic gastrostomy (PEG) or radiologically inserted gastrostomy (RIG) may be performed to bypass the oropharyngeal swallowing mechanism.

Currently, there is not clear evidence regarding the optimal timing for gastrostomy (H. Stavroulakis et al., 2014; T. Stavroulakis et al., 2013). However, due to surgical restrictions related to respiratory function, a gastrointestinal procedure may be recommended prophylactically. Current evidence suggests PEG procedures needs to be completed prior to a pwMND's functional vital capacity falling below 50% (Labra et al., 2020). RIG procedures require little, to no sedation and may be considered in pwMND who have impaired

respiration (McDermott & Shaw, 2008). Decision-making around gastrostomy is complex and multifactorial (Labra et al., 2020).

Factors that may contribute to a pwMND deciding to proceed with gastrostomy intervention include fatigue when eating, increased burden to prepare modified foods, increased risk of choking and aspiration and ongoing weight loss (H. Stavroulakis et al., 2014). Conversely, factors which may delay decision-making for gastrostomy include a reluctance to cease oral intake, negative perceptions and unclear benefits of gastrostomy and uncertainty of disease progression (H. Stavroulakis et al., 2014). For example, currently it is unclear if PEG tube placement is linked to extended survival (Sorenson et al., 2007) Notably, some pwMND have made the decision to undergo a PEG procedure, however experienced slower than anticipated swallowing function decline. Consequently, they decided to remove their feeding tube, to alleviate the associated discomfort and inconvenience of ongoing maintenance. Studies have reported that some pwMND may preference quality of life over length of life when deciding if to proceed with a PEG procedure, however what quantifies quality of life requires further exploration (Hogden et al., 2024). Additionally, pwMND may make decisions to proceed or refuse a PEG procedure based on their concerns of burdening family members (Hogden et al., 2024)

A pwMND and carers may be asked to participate in discussions about gastrostomy and make decisions about whether to proceed or refuse this intervention. Combining the above-described factors, decision-making related to gastrostomy is complex and requires cognitive engagement from a pwMND. Namely, a pwMND needs to understand the consequences of dysphagia as well as the concept and consequences of a medical procedure and ongoing enteral feeding. Additionally, these decision-making conversations require a pwMND to possess language capabilities, that facilitate their ability to voice their preferences.

Cognitive function in dysphagia management

Taken together, all of the assessment and management strategies for dysphagia in MND require a pwMND to cognitively engage. For example, during assessment, a pwMND will be asked to actively participate in discussions about their swallowing difficulties, which requires the ability to have insight and the ability to verbally express their experiences and views. Collectively, these discussions require a pwMND to first understand, then self-report on their level of function and then identify and describe their swallowing difficulty. In addition, swallowing trials and instrumental assessments require a pwMND to actively participate in and follow commands, for example to swallow on cue. Further, if dysphagia is subsequently identified, a pwMND will be provided with education and management strategies.

As described, dysphagia management often entails food modifications and safe swallowing strategies, as well as decision-making discussions regarding medical interventions such as enteral feeding. Collectively, these require cognitive engagement from a pwMND to enact recommended management strategies in the home environment, participate in discussions, and provide informed consent or refusal of interventions. However, there are complexities in dysphagia management in MND, as up to 50% of pwMND have co-occurring cognitive and behavioural changes (Abrahams, 2023; MND Australia, 2020a).

To support understanding of cognitive and behavioural changes in MND, typical cognition will be described below.

Cognition

In addition to motor control, the frontal lobe is involved in higher functions, or cognitive processes, such as executive function, attention, memory, language, social cognition, planning, inhibition and motivation (Fritz et al., 2014).

There are numerous broad definitions of what cognition is and what processes may be described as cognitive within the human species (Bayne et al., 2019). Essentially and simplistically, cognition can be conceptualised within domains of functioning (Harvey, 2019). The domains of cognitive function can be described within a hierarchical structure, with the bottom domains being associated with basic sensory and perceptual processing. Whereas the domains higher up refer to executive functioning and cognitive control. Therefore, cognition is a collective term that describes the intricate neural processes which allow humans to perceive, learn, remember, focus attention, make decisions, process and use language and plan meticulous sequences of motor movements (Bayne et al., 2019). These cognitive processes are essential for humans, who are social beings, to operate in a social environment, via complex intricacies of neural networks which allow humans to form relationships, understand the feelings and behaviours of others, and learn socialisation (Frith & Frith, 2012). Information, be it internal or external, is received, transformed, processed, saved and retrieved. The processing of this information allows humans to interpret the past, understand the present and predict the future, and to make sense of the surrounding environment. Consequently, cognitive processes are the foundation of human intelligence and behaviour (Harvey, 2019).

There is currently a lack of consensus as to which domains of functioning collectively delineate cognition, however, in the Diagnostic and Statistical Manual of Mental Disorders 5th edition (DSM-5), six principal domains (Figure 11) are described as: i. complex attention, ii. executive function, iii. learning and memory, iii. language, iv. perceptual motor function and v. social cognition.

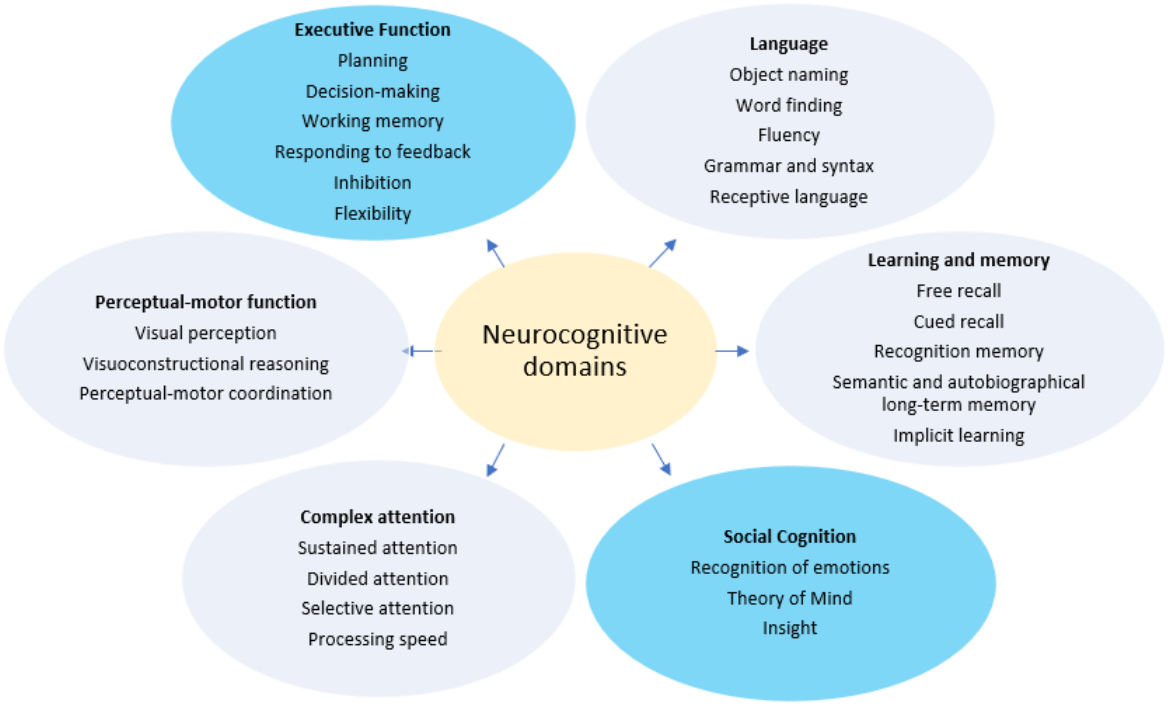

Figure 11. Neurocognitive domains as described in the DSM-5 (American Psychiatric Association, 2013) *(DSM-5: Diagnostic and Statistical Manual of Mental Disorders 5th edition).* Image by thesis author, adapted from American Psychiatric Association (2013)

Within this thesis, a focus will be placed on executive function and social cognition.

Executive Functioning

The first case study that described a connection between the frontal lobe and human personality involved railyard worker, Phineas Gage. Phineas sustained a traumatic workplace brain injury in September of 1848. Following an explosion, Phineas was struck by a 3.2cm diameter metal bar, penetrating his left cheek, irreparably damaging his left eye, as it passed through his skull and into the frontal lobes of his brain. Phineas survived his injury, however, was observed to experience significant changes to his personality (García-Molina, 2012; Teles, 2020). These changes manifested as reduced inhibitory control and an inability to regulate his behaviour. Consequently, Phineas' ability to function in his everyday life was severely impacted.

Figure 12: Phineas Gage. Image shows Gage holding the iron bar that injured him. Image retrieved from: *https://www.britannica.com/biography/Phineas-*

Whilst now considered to be medical folklore, the case study of Phineas Gage instigated neuropsychiatry investigations into the role of the frontal lobe. Specifically, theories about brain function localisation and the correlation with cognitive and behavioural sequelae followed, and links were established between the frontal cortex and executive functioning (García-Molina, 2012).

Executive functions (or executive functioning) relate to a complex collection of interrelated neurocognitive skills, critical for adaptive function (Diamond, 2013). Located within the neural networks of the prefrontal cortex, these processes allow both humans and animals to drive behaviour and execute goal-oriented tasks (Miyake et al., 2000). These top-down mental processes can be described as the separate functions of working memory, inhibition, cognitive flexibility and interreference control (Miyake et al., 2000). Planning, decision making, making mental adjustments to information and adjusting behaviours within different contexts are all actions which draw on these cognitive processes (Diamond, 2013; Miyake et al., 2000).

Typical executive functioning is dependent on neural networks within both the prefrontal and parietal cortex, basal ganglia, thalamus and cerebellum (Kim et al., 2017; Nowrangi et al., 2014). Consequently, any change to the neural networks which may occur as a result of disease or trauma, can manifest as changes to executive functions.

Social Cognition

Social cognition involves various neurobiological processes which underpin effective social behaviour and interactions (Elamin et al., 2012). Humans are a social species, and it is these social cognitive processes which enable humans to be a part of social groups, to both connect and to learn. Social cognition develops from infancy and becomes more automatic in adulthood (Frith & Frith, 2012). For example, social referencing begins to develop as infants observe and learn from their carer's facial expressions, helping them to determine when things are safe or unsafe (Frith & Frith, 2012). Additionally, as adults, humans automatically focus attention and look towards something, purely because another person is looking in that direction, in a social process known as gaze following (Frith & Frith, 2012). These social cognitive abilities, which allow for the analysis of facial expressions and eye movements are associated with the posterior region of the superior temporal sulcus and the fusiform gyrus (Frith & Frith, 2012). Further, emotional contagion is a basic conditioning mechanism, which enables humans to both recognise and share the emotions felt by others, through perception. For example, when someone smiles, the natural human reaction is to smile back, automatically aligning with the emotions of the other person. Emotional contagion is said to be a prerequisite process for developing and displaying empathy (Frith & Frith, 2012) and contributes to Theory of Mind.

Theory of Mind (ToM) or mentalising, is a meta cognitive process, associated with regions in the medial pre-frontal cortex (Frith & Frith, 2012). ToM enables humans to make inferences about the internal processes of self and others, specifically the internal emotions, intentions and beliefs, that cannot be directly observed. It allows for humans to reflect on the actions of

self and other's and to think about one's own thoughts. Further, ToM allows humans to both connect socially and understand and predict the behaviour of others (Frith, 2008). Theory of Mind can be further divided into cognitive and affective ToM. Cognitive ToM can be described as knowing how something will be received by another, and affective ToM can be described as knowing how someone will feel (Elamin et al., 2012).

There are many processes, which happen both subconsciously and automatically, that contribute to social cognition, including memory, perception, attention and planning, as well as the higher order social concepts such as irony, sarcasm and humour. Social cognition may also be referred to as information processing, consisting of a stimulus and a response (Elamin et al., 2012). Within this, there is a subset of processes: the perception of a stimulus, decisions about what to do about the stimulus and then a social response.

The neural substrates of social cognition include the amygdala, the orbitofrontal cortex, the ventromedial prefrontal cortex and the temporal poles, predominantly of the right hemisphere of the brain (Adolphs, 2009; Elamin et al., 2012; Frith, 2008). Part of the limbic system, the amygdala, is an almond shaped structure located within the temporal lobe. The amygdala functions to control emotions and is understood to regulate anxiety, fear conditioning, aggression and social cognition (AbuHasan et al., 2023). Along with the orbitofrontal and anterior temporal cortex, and the thalamus, the amygdala forms part of the basolateral circuit, believed to be the substrate of social cognition (AbuHasan et al., 2023). Many social processes of social cognition occur automatically and without conscious awareness. However, executive control of social cognition is an important factor, and it has been shown that areas of the dorsolateral prefrontal cortex function to influence over the amygdala, modulating the response (Frith & Frith, 2012).

When progressive neurodegeneration occurs to areas within the pre-frontal cortex or temporal lobes of the brain, this may present clinically as progressive deficits in executive

functioning, changes to behaviour or language impairments (Bang et al., 2015). With continued decline, a person may be diagnosed with frontotemporal dementia.

Frontotemporal dementia

Frontotemporal dementias (FTD) are a group of neurodegenerative diseases, which clinically present as changes to personality and social behaviours (Yancopoulou et al., 2003). Also referred to as frontotemporal lobar degeneration (FTLD), FTD is clinically and pathologically heterogeneous, and is characterised by neuronal loss, synapse loss, and gliosis (Ljubenkov & Miller, 2016) in the frontal lobe and temporal lobes. This neuronal loss, consequently, leads to gross atrophy within the frontal and anterior temporal lobes, basal ganglia and thalamus (Ljubenkov & Miller, 2016; Rabinovici & Miller, 2010). FTD is a rare disease, affecting 15-22 per 100,000 people in Australia (Ratnavalli et al., 2002). However, FTD is the most common dementia in younger people with symptom onset typically occurring between the age of 45 to 65 years (Ljubenkov & Miller, 2016). up to 40% of FTD cases are familial (De Marchi et al., 2023).

FTD differs from other forms of dementia, such as Alzheimer's disease. Pathologically, in Alzheimer's disease, there is a formation of beta amyloid plaques (protein fragments, which become sticky and clump to form plaque). It is currently unclear if these plaques contribute to, and are therefore, the cause of Alzheimer's disease, or if the plaques form as a consequence of the disease (Scheltens et al., 2021).

FTD however, is known to be associated with pathogenic variants of three genes, specifically the most common variant, C9orf72 (as described in the genetics in MND section above), progranulin (GRN), and microtubule associated protein tau (MAPT) associated with the neurofibrillary tangling of proteins (Jellinger & Attems, 2007). Specifically, in FTD, the pathological proteins include the TAR DAN binding protein (TPD-43), the microtubule associated protein tau protein and in rarer cases the heterogeneous nuclear ribonucleoprotein P2 or FUS protein (Figure 13).

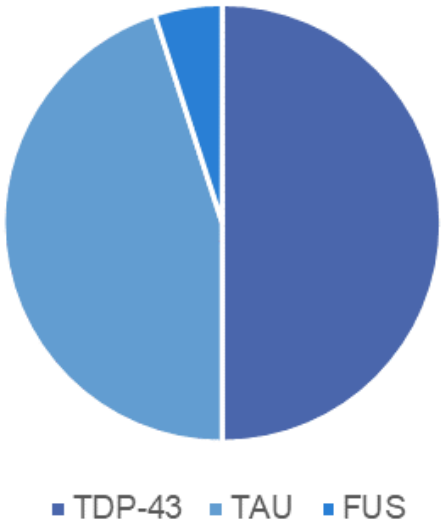

Figure 13: Protein pathologies associated with fronto-temporal dementia. TDP-43, TAU and FUS proteins
(TDP-43: TAR DAN binding protein; TAU: microtubule associated protein; FUS: heterogeneous nuclear ribonucleoprotein P2). **Image created by thesis author.**

FTD can be further sub-categorised based on the clinical presentation as behavioural-dominant FTD or language-dominant FTD (Rabinovici & Miller, 2010). Behavioural dominant FTD corresponds with neuronal loss specific to the frontal cortex and anterior parts of the temporal lobe (Strong et al., 2009). The language dominant FTD is also referred to a primary progressive aphasia and can be further sub-categorised as shown in Table 2. Specifically, patterns of neuronal loss in a language-dominant FTD reveal more severe lateral inferior temporal lobe degeneration (Coon et al., 2011). As language is known to be lateralised to the left temporal lobe, this clinical presentation of a language-dominate FTD is expected.

Table 2.

The subcategories of frontotemporal dementia with description of clinical presentation and pathophysiology

FTD type	Clinical presentation	Pathophysiology
Behavioural variant FTD (bvFTD)	Changes in personality and behaviour. Apathy and disinhibition. Reduced empathy and insight	TAR DAN binding protein (TDP-43) Frontal cortical degeneration (Rabinovici & Miller, 2010)
Language dominant FTD or Progressive primary aphasia (PPA)	Word finding difficulties. Impaired people and object naming,	
Progressive non-fluent aphasia (also referred to as agrammatic PPA)	Slow effortful speech with impairments in both speech production and grammar	Predominant left posterior frontoinsular atrophy +/- hypometabolism (Kiymaz et al.; Rabinovici & Miller, 2010)
Sematic dementia or semantic PPA	A progressive loss of 'semantic' knowledge about words, objects and concepts.	Disrupted network connectivity within the left dominant anterior temporal region. TDP-43 pathology (Kiymaz et al.)
Logopenic PPA variant	Word finding difficulties	Pre-cursor to Alzheimer's disease pathology in 95% of cases. TDP43 pathology less commonly reported. Left posterior, superior and middle temporal gyri and inferior parietal lobe (Kiymaz et al.; Rabinovici & Miller, 2010)

In addition to being associated with changes to personality, social behaviour and language, FTD is associated with a wide range of motors signs, including those commonly observed in Parkinson's Disease, Supranuclear Palsy, Corticobasal Syndrome and MND (Schönecker et

al., 2022). Notably, approximately 15% of people with a confirmed FTD diagnosis will progress to develop motor signs (Ljubenkov & Miller, 2016). Additionally, impairments in frontotemporal functions, which may manifest as a spectrum of clinical deficits to both cognition and behaviour (Strong et al., 2009) may present in some who receive an MND diagnosis.

Cognition in MND

Reports of non-motor symptoms in MND have long been reported, dating back to the late 1880s, however cognitive and behavioural descriptions used in early writing differed substantially to present day, with terms such as “mentally feeble”, “delusional”, “paranoid” and “schizophrenia” used (Bak & Hodges, 2001). Additionally, most of these original findings were not published in English and consequently the association between cognitive and behavioural changes in MND remained largely unexplored until the 1980s (Hudson, 1981). Later, in 2002, a cross-sectional study investigated the co-association of FTD and MND (within an FTD population) (Lomen-Hoerth et al., 2002). Of the 36 participants with a confirmed FTD diagnosis, 14% were also found to have a definite MND diagnosis, and an additional 36% of these received a probable MND diagnosis. These findings confirmed that neuromuscular characteristics were present in an FTD population, and additionally, suggested an overlap between FTD and MND (Lomen-Hoerth et al., 2002). However, it was rare at this time for people, with a diagnosis of MND, to undergo neuropsychological testing, given it was seldom considered that there may be a co-occurrence of dementia-like symptoms (Woolley & Strong, 2015). Further, the loss of motor function and communication impairments in MND makes identification of cognitive and behavioural deficits more difficult (Bak, 2010).

In around 2006, after continued investigations, the TAR-DNA binding protein (TDP-43) was discovered to be the pathological protein associated with both MND and FTD (Kiernan et al., 2011; Lomen-Hoerth et al., 2002b; Neumann et al., 2006). This finding confirmed the

existence of an FTD continuum in MND, and that cognitive and behavioural changes are associated with MND.

Ongoing research led to the 2011 discovery of the C9orf72 genetic expansion occurring on chromosome 9p21.1. It was this discovery that provided the most convincing genetic evidence between the overlap of FTD and MND (Gijselinck et al., 2018; Renton et al., 2011). Since then, the defective C9orf72 repeat expansion gene (of a GGGGCC intronic repeat sequence) has been found to occur in approximately 30% of familial MND cases and approximately 10% of sporadic onset MND cases (Kumar et al., 2016). In people with familial MND, the C9orf72 mutation also reveals a TDP-43 aggregate pathology as described above. Clinically, this pathology is associated with cognitive and behavioural changes.

Similarly to FTD, cognitive and behavioural changes in MND are also further sub-categorised into behavioural-dominant symptoms and language-dominant symptoms (Coon et al., 2011). Post-mortem studies reveal frontotemporal lobar degeneration (Neary et al., 2000), with patterns of neuronal degeneration in behavioural FTD MND more localised to the frontal lobe, whereas language-dominant FTD MND show more severe loss in the lateral inferior temporal lobes (Coon et al., 2011). Behavioural-dominant FTD MND is associated with longer survival rates than the language-dominant FTD MND variant (Coon et al., 2011). Additionally, there is an association between language-dominant FTD MND and bulbar onset MND. Consequently, cognitive and behavioural changes are reported to occur in up to 50% of pwMND, with up to 15% meeting the diagnostic criteria for FTD, usually presenting as a behavioural variant FTD (Abrahams, 2023; Abrahams et al., 1996; Kew & Leigh, 1992; Pender et al., 2020; Strong et al., 2017).

Cognitive change in MND prominently manifests as executive dysfunction (Goldstein & Abrahams, 2013; Pender et al., 2020; Phukan et al., 2007), with 35% of pwMND reported to experience mild to moderate cognitive impairment which typically occurs early in diagnosis

(Pender et al., 2020). Further, as MND progresses, this coincides with increased severity of neuropsychological dysfunction (Pender et al., 2020). The neuropsychological deficits in MND are heterogenous and linked to poorer patient outcomes and reduced survival (Strong et al., 2017). Additionally, pwMND assessed to be cognitively unaffected at baseline have been shown to develop cognitive dysfunction as the disease progresses (Pender et al., 2020).

A sensitive clinical marker to evaluate executive function in MND is via verbal fluency assessment (Strong et al., 2017). In clinical assessment, verbal fluency in MND is impaired, and subsequent neuroimaging reveals frontal lobe dysfunction, specifically in the dorsolateral prefrontal cortex and inferior frontal gyrus (Strong et al., 2017). In addition to executive function deficits in MND, social cognition is impaired (Goldstein & Abrahams, 2013).

People with MND have been shown to have deficits in social cognition related to impaired emotional processing, an impaired ability to recognise emotional facial expression, difficulty understanding social situations and impaired ToM (Strong et al., 2017). A broad range of behavioural descriptors have been used as associated with behavioural changes in MND (Figure 13), which are described within seven subcategories i. disinhibition, ii. apathy, iii. perseveration and/or compulsive behaviour, iv. hyperorality, v. loss of sympathy/empathy, vi. psychotic symptoms, vii. loss of insight (Trucco et al., 2024). These descriptors for cognitive and behavioural changes in MND mostly correspond with the behavioural descriptors used in behavioural-variant FTD (Trucco et al., 2024).

Image removed due to copyright

Figure 14: Behavioural symptoms associated with MND with and without FTD. As described in Trucco et al. (2024)
(MND: Motor Neurone Disease; FTD: Frontotemporal dementia)

Further, investigations via computed tomography (CT) and electroencephalogram (EEG) assessment show that pathological cortical and neuronal changes can occur before cognitive and behavioural changes manifest clinically (Gallassi et al., 1989).

There is increasing attention within the scientific community about cognitive and behavioural changes in MND. This attention led to the revised Strong criteria being published in 2017 (Strong et al., 2017), after acknowledgement that the initial Strong criteria, published in 2009 did not recognise impairments to executive functions, language or social cognition (Strong et al., 2017). However, despite this advancement in knowledge of cognitive and behavioural changes as being associated with MND, currently, there are still misconceptions both within healthcare and the broader public community that cognition in MND remains spared.

In summary, cognitive and behavioural changes in MND may occur prior to motor impairments, cooccur with motor symptom onset, or develop as the disease progresses. Earlier clinical presentations of cognitive and behavioural changes in MND are associated with poorer prognosis and reduced survival. Whilst there is increasing scientific attention to cognitive and behavioural changes in MND, as yet our understanding is incomplete, and within the broader community, the association between cognitive and behavioural changes in MND is not widely known.

Additional non-motor signs of MND

In addition to the non-motor symptoms of cognitive and behavioural changes outlined above, an increasing body of literature is now exploring other non-motor symptoms of MND. For example, whilst sensory changes are not thought to be associated with MND, emerging evidence increasingly demonstrates the contrary (Isaacs et al., 2007; Rubio et al., 2022).

Sensory neuropathy has been documented in pwMND, with sensory nerve biopsies revealing sensory axonal dysfunction, (Isaacs et al., 2007) with axonal changes reported to progress over time (Rubio et al., 2022).[1]

As previously described, swallowing is a sensorimotor process, reliant on sensory input to modulate appropriate motor output. As such, it is important to consider alterations to sensation in MND in the context of dysphagia.

Recently, Paterson, Doeltgen, Francis (2024), synthesised in their scoping review, evidence of sensory changes that are related to swallowing function. Evidence for changes to olfactory perception, laryngeal, palatal and pharyngeal sensation, altered taste and smell, and changes to cough reflex were reported. Of note, the available descriptions were varied and inconsistent, with most studies describing decreased sensory changes. Overall, however,

[1] Whilst sensory change is not a key aspect of the aim of this program of research, this summary section is provided for contemporary context.

the evidence base for swallowing-related sensory changes in MND is weak and lacks methodological rigour. However, given that pwMND experience silent aspiration, and anecdotally report changes to appetite, olfaction and taste, combined with evidence of changes to sensation occurring in FTD, more work is warranted in this space.

MND Management

As there is no cure for MND and few pharmalogical therapies currently available, treatment of MND consists mainly of symptom management and palliative care approaches (McDermott & Shaw, 2008). Specialised multidisciplinary teams (MDT) have been shown to improve patient outcomes in MND (Traynor et al., 2003). It is recommended that these specialist teams consist of a neurologist, physiotherapist, respiratory physician and respiratory nurse, specialist nurse, social worker, dietician, speech pathologist and palliative care doctors (McDermott & Shaw, 2008; National Institute for Health and Care Excellence, 2016). MND care is described as specialised, and MDT are to ensure effective communication and coordination amongst the team, with the provision of regular and repeated assessments for all symptoms of MND. Further the International Alliance of ALS/MND describes best practice principles for MND care.

According to the International Alliance of ALS/MND Associations statement (MND Australia, 2010), good practice for the management of MND is underpinned by the following five principles:

1. Management of the disease is determined by the needs and wishes of the person living with ALS/MND, treating the person with ALS/MND with care, respect and dignity
2. Timely response to identified needs
3. Access to a coordinated and integrated care plan
4. Regular monitoring and review of the person's condition, and appropriateness of the care plan
5. Information about the person's medical condition held in confidence

Specifically, this statement describes that at the time of MND diagnosis, families are provided with both verbal and written information about the disease to support both their

understanding and the impact of MND. It is also recommended that the provision of information to families continues throughout the entirety of a families' MND care journey.

Palliative care referrals are suggested at the time of diagnosis of MND (Andersen et al., 2012; Andersen et al., 2005). As there are no current treatments available for MND, palliative care plans aim to maximise a pwMND's quality of life and manage presenting symptoms to optimise comfort. Additionally, it is recommended that emotional and psychological support is offered through palliative care teams. Through these support services, discussions regarding advanced care and end of life preferences are also recommended.

The NICE guidelines for assessment and management of MND (National Institute for Health and Care Excellence, 2016) alert HCPs to cognitive changes and FTD as cooccurring in MND. This document recommends that cognitive assessments are undertaken and that care for a pwMND, who has cognitive and behavioural changes, is provided in accordance with the Mental Capacity Act 2005 (Office of the Public Guardian & Ministry of Justice, 2023). Within this act, a person who is deemed to lack capacity is defined as "…a person who lacks capacity to make a particular decision or take a particular action for themselves at the time the decision or action needs to be taken." (Office of the Public Guardian & Ministry of Justice, 2023, p. 3). The NICE guidelines also describe tailoring all conversations to a pwMND's capacity.

Additionally, and interestingly, within the NICE guidelines, dysphagia is not described as a separate symptom of MND, but rather as a consideration within Nutrition and Gastrostomy management (National Institute for Health and Care Excellence, 2016). Here, reference to cognitive changes is also mentioned, stating that a person's capacity to make decisions about medical care in relation to nutrition and gastrostomy needs to be determined. However, specific strategies to guide HCPs to navigate this are not provided in these guidelines.

Navigating MND management is complex, and for MDTs to be effective, care needs to be coordinated to allow all members of the team to collaborate together, with efficient methods of communication and opportunities to debrief and discuss all facets of care. The focus of MND management should be to support patient autonomy and provide support to families as they come to terms with an MND diagnosis, as well as severe changes to motor function (Leigh et al., 2003). As also highlighted, MND is now recognised as a multisystem disease, negatively impacting on cognition, behaviour and language in some.

As part of MND management, cognitive and behavioural changes need to be considered, and are an important aspect both in general and more specifically in dysphagia management. However, this does not appear to always take place. Hence it is important to evaluate this.

Therefore, Chapter 2, describes the specific studies and the study aims that were addressed and contribute to answering the overall research question.

CHAPTER 2: RESEARCH AIMS AND OBJECTIVES

Overall research aims and objectives

The overarching aim of this program of research is to understand how cognitive and behavioural changes in MND interact or impact on dysphagia management. The additional aims of this research include i. to understand management approaches to cognitive and behavioural changes in MND more broadly and ii. understand the preferences of families living with MND for both dysphagia and cognitive and behavioural changes.

To address these aims, the following research questions were explored*:*

1. How do cognitive and behavioural changes impact on dysphagia? (Chapter 3)
2. How are cognitive and behavioural changes in MND managed? (Chapter 5)
3. How do HCPs recognise, provide information on, and manage cognitive and behavioural changes in MND, and what enablers and barriers are there to this? (Chapter 6 and 7)
4. What are the factors that influence clinical management of cognitive and behavioural changes in MND? (Chapter 6 and 7)
5. What are the experiences of people living with MND, their carers and family members of understanding and managing dysphagia? (Chapter 8)
6. What are the facilitators and barriers for people living with MND for implementing and/or adhering to a swallowing management plan? (Chapter 8)
7. How do cognitive and behavioural changes impact on pwMND's understanding and management of their swallowing impairment? (Chapter 8)
8. How do pwMND & carers experience the symptoms of dysphagia and cognitive and behavioural changes in MND, and what are their preferences for management. (Chapter 8)

The next section outlines the research aims, questions, rationale, and significance of the six research studies undertaken and presented in this thesis.

Research project 1 - The impact of cognitive decline in MND on swallowing. A scoping review (Chapter 3)

Aims

This study aimed to identify what is known about cognitive and behavioural changes in MND and how these changes interact with dysphagia management, and summarise the literature related to swallowing and cognitive and behavioural changes in MND.

Research Question

How does cognitive impairment impact on a pwMND's ability to understand and manage oropharyngeal swallowing function?

Rationale and Significance

Cognitive and behavioural changes are associated with MND; however, it is currently not known how these changes interact with dysphagia management. Understanding how cognitive and behavioural changes in MND affects how a pwMND understands and manages dysphagia is important because these changes have been shown to impact on a pwMND's ability to understand complex information, their decision-making capacity (Khin Khin, Minor, Holloway, & Pelleg, 2015) and flexibility. As oropharyngeal swallowing assessment and management require a pwMND to actively participate and enact, cognitive engagement is required. Dysphagia in MND is associated with poorer outcomes, and it is critical to understand if the added presence of cognitive and behavioural changes further complicates swallowing management and outcomes (Paris et al., 2013).

Research project 2 - How do people living with MND understand and manage a swallowing disorder? An ethnographic study (Chapter 4)

Aims

This study aimed to explore how pwMND, who may or may not experience cognitive and behavioural changes, their primary carers, manage swallowing function outside of the clinic context. An additional aim was to explore how swallowing management plans, such as those recommended by speech pathologists, are enacted in the home environment.

Research Questions

1. What are the experiences of people living with MND, their carers and family members of understanding and managing dysphagia?
2. What are the facilitators and barriers for people living with MND for implementing and/or adhering to a swallowing management plan?
3. How do cognitive and behavioural changes impact on pwMND understanding and management of their swallowing impairment?

Rationale and Significance

PwMND and their carers often need to make changes to how they manage their swallowing (dys)function as the disease progresses. Some pwMND may also experience cognitive and behavioural changes, in addition to dysphagia. Currently, it is not understood how pwMND and their carers manage swallowing changes in the home environment. In addition, it has also not yet been evaluated how cognitive and behavioural changes may interact with dysphagia management strategies. Currently, there is no information that tells us how cognitive and behavioural changes impact on how a swallowing problem might be understood or managed.

Understanding this interaction is important to learn how pwMND, their carers and families with/without cognitive and behavioural changes make informed decisions on dysphagia

management, and implement safe swallowing strategies in the home, to support adequate nutrition and hydration. This information is also important to inform clinical practice and improve patient care.

Research project 3 - Exploring clinical management of cognitive and behavioural deficits in MND. A scoping review (Chapter 5)

Aims

This study aimed to identify and collate strategies specific to managing cognitive and behavioural changes in MND.

Research Question

How are cognitive and behavioural changes associated with MND managed?

Rationale and Significance

Management of cognition and behaviour for pwMND has critical implications for both supporting people to participate in decision-making regarding their disease and being able to functionally communicate their preferences and decisions. However, currently, there is little empirical evidence that exists to guide families living with MND and HCPs. Collating the available evidence of management strategies for cognitive and behavioural changes in MND provides a valuable resource for HCPs. Additionally, it highlights areas that require continued investigation to improve clinical practice and care for pwMND.

Research project 4 - A Survey of HCPs supporting people with MND (Chapter 6)

Aims

This study aimed to better understand how HCPs working with pwMND and their families currently receive training on, recognise, provide education for, and subsequently manage, cognitive and behavioural changes in MND. Additionally, the aim was to identify facilitators and barriers to managing cognitive and behavioural changes in MND.

Research Question

How do HCPs recognise, provide information on, and manage, cognitive and behavioural changes in MND, and what enablers and barriers are there to this?

Rationale and Significance

It is important to understand the extent that cognitive and behavioural changes are being addressed in clinical practice and identify any barriers that exist to both assessing and managing cognitive and behavioural changes in MND. This understanding will help to inform and design clinical pathways, professional development opportunities and future research to underpin better evidence-based practice.

Research project 5 - Understanding practice: Factors influencing management of cognitive and behavioural changes in MND. A qualitative study using the Theoretical Domains Framework (Chapter 7)

Aims

This study aimed to explore HCP's experiences of addressing and managing cognitive and behavioural changes in MND, including individual factors which influence clinical approaches to identifying and managing cognitive and behavioural change in MND. Further, this study aimed to explore the potential facilitators and barriers to management of cognitive and behavioural changes in MND, as well as HCPs behaviours and attitudes related to their clinical practice

Research Question

What factors influence management of cognitive and behavioural changes in MND?

Rationale and Significance

Detection and the subsequent management of cognitive and behavioural change in MND is important, as these changes are associated with negative outcomes for families living with MND related to reduced treatment uptake, reduced treatment adherence (Hill, 2016; N. H. Martin et al., 2014), reduced quality of life for pwMND (Radakovic et al., 2024) and increased caregiver burden (Pagnini et al., 2010).

Research project 6 - What are families' experiences of dysphagia and cognitive and behavioural changes in MND? What are their preferences for care? (Chapter 8)

Aims

This study aimed to explore the lived experience of both dysphagia and cognitive and behavioural changes in MND for both pwMND and their carers, as well as exploring families' preferences for care related to both dysphagia and cognitive and behavioural changes in MND.

Research Questions

1. How do pwMND & carers experience the symptoms of dysphagia and cognitive and behavioural changes in MND?
2. What are their preferences for management?
3. How do pwMND & carers experience the symptoms of dysphagia and cognitive and behavioural changes in MND, and what are their preferences for management.

Rationale and Significance

Almost all pwMND will experience dysphagia as the disease progresses (Waito et al., 2017), significantly affecting health and quality of life (Paris et al., 2013). In addition, some pwMND experience cognitive and behavioural changes, which have been shown to also impact on a pwMND's quality of life and is linked to increased carer burden (de Wit et al., 2018). However, currently it is not yet known how families with MND experience dysphagia, or cognitive and behavioural changes in MND, or what their preferences are for the management of these symptoms. Learning about the preferences of families with MND to manage cognitive and behavioural will support the development of clinical frameworks and resources to guide HCPs. This will hopefully lead to better outcomes and improve care outcomes for families with MND.

Methodology and methods

The overarching methodology underlying this program of research is situated in constructivism.

Ontology can be described as the study of existence or being, and the nature of reality (Guarino et al., 2009). The ontological perspective of constructivism proposes that the social phenomenon of knowledge is actively constructed and shaped through experiences, human interactions and reflection, rather than being passively absorbed (Adom et al., 2016; Schwandt, 1994). Constructivism is in opposition with positivism, which suggests that there is only one reality that exists, which is to be discovered as objective knowledge. In positivism's objective stance, it positions the world as measurable. (Adom et al., 2016; Alharahsheh & Pius, 2020). This is in contrast to constructivism, which takes an interpretivist stance, that does not purport to measure meaning. In constructivism, learning is a process of constructing meaning through interactions within an individual's environment and an individual's reality of experiencing and doing. Constructivism also entails that the acquisition of new knowledge is dependent on prior knowledge and experiences, and this prior knowledge influences and informs how an individual acquires new knowledge (Amineh & Asl, 2015).

Epistemology, founded in the Greek words of *episteme* meaning "knowledge" and *logos* meaning "argument", refers to the processes by which knowledge is gained and how we justify our beliefs of knowledge (Alharahsheh & Pius, 2020; Steup & Neta, 2005). The epistemological approach for this program of research is grounded in both constructivist and interpretivist paradigms, which posits that knowledge is constructed, interpretative and unique in nature and its meaning needs to be explained (Alharahsheh & Pius, 2020).

Consequently, with a constructivist approach to enquiry, researchers consider many factors, such as the interplay of social relationships between the phenomena, to develop knowledge (Pouliot, 2004). For example, the communities, societies, cultures and institutes to which

people belong and interact with may impact on their individual reality of any given situation (Creswell, 2008; Pouliot, 2004). In this context, this program of research acknowledges the role that subjective experience has on the formation of an individual's reality. Consequently, it is the interaction with these external factors that create an individual's reality, and it is within these varied and nuanced perspectives of one's reality that enriched knowledge of a phenomenon may be developed (Amineh & Asl, 2015; Pouliot, 2004).

Therefore, using a constructivist approach allowed the researcher to construct meaning about the interaction between dysphagia and cognitive and behavioural changes in MND through investigations, that consider the experiences of families living with MND and the HCPs who support pwMND. Taking a constructivist standpoint for this program of research further enabled the use of qualitative methods that allow for an in-depth exploration of the interaction between dysphagia and cognitive and behavioural changes in MND.

As such, the qualitative approaches undertaken in this program of research are situated within constructivist ontology and interpretivism (Alharahsheh & Pius, 2020). Data analysis within constructivist research is interpretative and iterative, and subsequently the researchers undertook a continual process of refinement of understanding as new insights emerged.

First, a scoping review was conducted to explore and synthesise the literature, which considered dysphagia and cognitive and behavioural changes in MND (Chapter 3) applying the framework described by Arksey and O'Malley (2005). This rigorous and systematic approach allowed the researcher to scope the breadth of the published peer reviewed literature as well as the publicly available information, relevant to the research question. To analyse these data, we employed thematic analysis, using the methods described by Braun and Clarke (Braun & Clarke, 2006), which acknowledges the importance of recurrence and pattern making within the data, however, also allows the researcher to construct meaning

and meaningfulness throughout the coding processes. This allowed the researcher to identify patterns and unique interpretations in these data.

Chapter 4 describes a planned ethnographic study. Ethnography is underpinned by constructivism and involves the exploration of people and culture, aiming to capture the complexity of a participant's social life and social connections (Goodson & Vassar, 2011). An ethnographic approach to research involves consideration of the viewpoint of the participant, with a focus on lived experience and social patterns, within a participants natural setting (Goodson & Vassar, 2011). As such, we aimed to explore the experiences of families with MND who have dysphagia with, or without, cooccurring cognitive and behaviour changes, within their home environment. As described in Chapter 4, this study was impacted by Covid-19 and was not able to be completed.

Due to the global pandemic, a pivot to this program of research was required (as shown in Figure 1). The findings of the first scoping review (Chapter 3) revealed a gap in knowledge related to the interaction between dysphagia and cognitive and behavioural changes in MND. As such a second scoping review was undertaken (Chapter 5), which aimed to understand management approaches to cognitive and behavioural changes in MND more broadly. To this end, the same methodology as described in Chapter 3 was followed, with a systematic scoping review of the literature (Arksey & O'Malley, 2005) and thematic analysis of the data conducted (Braun & Clarke, 2006).

Chapters 6 describes a mixed methods study aimed at understanding how HCPs manage cognitive and behavioural changes in MND. Specifically, this research project consisted of a concurrent mixed methods survey, that combined aspects of both quantitative and qualitative research (Creswell, 2008, 2014).

The concurrent mixed methods survey design (Chapter 6) allowed for quantitative and qualitative data collection to occur at the same timepoint, within the same survey project.

Data analysis of the survey then consisted of an integration of the quantitative and qualitative data to provide a comprehensive descriptive analysis of these data (Creswell, 2008), describing frequency, experiences, attitudes and beliefs related to cognitive and behavioural changes in MND. The Theoretical Domains Framework (TDF) (Cane et al., 2012) is an implementation and behaviour change framework tool which may be applied to understand and/or explain factors that influence implementation. Additionally, the TDF framework enables identification of characteristics of behaviour that may require change.

We focussed the TDF both in relation to current HCPs practices (Chapter 6 and Chapter 7) and to pwMND and carers' experiences (Chapter 8) to allow for in depth analysis of behaviour change and implementation science in relation what is currently being done and what is not being done in clinical practice for cognitive and behavioural changes in MND. Consideration of implementation science is essential when attempting to create any type of behaviour/practice change. The TDF framework enabled us to identify and understand the knowledge, skills and attributes of current practices and to identify where the gaps in current practices are. There is flexibility in the application of the TDF framework; and within Chapter 6 it was applied as an interpretative framework to assist in understanding the facilitators and barriers to addressing and managing cognitive and behavioural changes in MND.

Following this, participants, who agreed to partake in the survey were then invited to participate in qualitative semi-structured interviews as presented in Chapter 7. These semi-structured interviews were conducted to gather HCPs' personal experiences, perspectives, and attitudes and to provide a deeper understanding of how HCPs approach cognitive and behavioural changes in MND. The interview guide was developed, based on the findings of the survey and was iteratively refined, based on consultation with the research team. These data were then analysed, first deductively to the domains of the TDF, and then inductive coding was conducted to develop categories. Finally, thematic analysis was conducted to identify themes (Braun & Clarke, 2006).

To further explore the research question and to learn from families who are living with MND, qualitative semi-structured interviews were conducted with both pwMND and their carers (Chapter 8). These interviews were conducted to better understand the thoughts, feelings, experiences and preferences of both pwMND and carers, in relation to dysphagia and cognitive and behavioural changes in MND. The focus of the interactions within this study were between the pwMND and HCPs and the carers and HCPS. Whilst the interaction between pwMND and their carers is recognised as very important, it is outside this program of research. A semi-structured interview guide was developed and iteratively refined based on consultation with the research team. To support data analysis, the TDF was used, with initial coding completed deductively to each TDF domain. Then, to further categorise these data within each domain, we conducted inductive coding to develop categories. Thematic analysis (Braun & Clarke, 2006) was then undertaken to identify themes within this data set.

Ethics approval

Ethics approval was granted by the Human Research Ethics Committee at Flinders University. Ethics Approval: number 4660 (Appendix 1) relates to Chapter 6, 7 and 8 which involved participants. All participants were provided with a participant information sheet and consent form. Consent for online interviews was gained verbally prior to starting the interview. In addition, Ethics Approval 2850 relates to Chapter 4 (Appendix 2).

In summary, this program of research is grounded in constructivism in that it is exploratory, designed to synthesise the literature and explore the participant's experiences, beliefs and perceptions.

> *How do cognitive and behavioural changes in MND interact with dysphagia management?*

The findings from each study as they relate to the above research questions will be explored in the Chapters within Part 2.

Part 2

CHAPTER 3: THE IMPACT OF COGNITIVE DECLINE IN MND ON SWALLOWING. A SCOPING REVIEW

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Impaired swallowing is a serious symptom of MND impacting on health and wellbeing. Little is known about how cognitive impairment in MND impacts on oropharyngeal swallowing. A scoping review was undertaken to address the following research question:

How does cognitive impairment impact on a person living with MND's ability to understand and manage oropharyngeal swallowing function? [2][3][4]

The paper was co-authored, and the contribution of each author was as follows:

Ms. Rebecca Francis
Research design, 60%, Data collection and analysis 70%, Writing and editing 70%

Associate Professor Stacie Attrill
Research design, 20%, Data collection and analysis 15%, Writing and editing 15%

Professor Sebastian Doeltgen
Research design, 20%, Data collection and analysis 15%, Writing and editing 15%

[2] Note the published version of this study used the acronym ALS, however for the purpose of this thesis ALS has been amended to MND where possible.

[3] The chapter was peer reviewed and published online in the International Journal of Speech-Language Pathology in 2021, and subsequently in print.

[4] References within this chapter have been reformatted to meet the requirements of this thesis.

Introduction

Almost all pwMND develop an oropharyngeal swallowing impairment over disease progression (Burgos et al., 2018; Murono et al., 2015; Onesti et al., 2017) and increasing research suggests that cognitive impairment also occurs in up to 50% of pwMND (Chiò et al., 2012; Flaherty-Craig et al., 2011; Portet et al., 2001; Simon & Goldstein, 2019). However, little is known about the impact of cognitive impairment on how pwMND understand and manage oropharyngeal swallowing function. This is important because cognitive impairment is associated with reduced capacity for decision-making, treatment uptake and adherence (Chiò et al., 2012; Flaherty-Craig et al., 2011; Khin Khin et al., 2015). Studies that explore the relationship between oropharyngeal swallowing function and cognitive impairment associated with MND are necessary to determine how this affects the way people experience swallowing, make informed decisions on swallowing management, and adhere to safe swallowing recommendations.

Impaired oropharyngeal swallowing in MND increases the risk of malnutrition, dehydration, and aspiration (Ruoppolo et al., 2013), negatively affects quality of life (Vesey, 2017) and increases caregiver burden (Aoun et al., 2013; Geng et al., 2017; Ng et al., 2011). The main cause of death in MND is related to respiratory failure (Govaarts et al., 2016; Vesey, 2017), and oropharyngeal swallowing impairments compromise an already at-risk respiratory system. In addition, pwMND experience anxiety and fear related to choking (Oliver & Turner, 2010). Therefore, pro-active and well-informed swallowing management is critical. As the disease is terminal and often rapidly progressing, interventions prioritise prolonging survival and managing symptoms to improve function and quality of life. However, there is increasing awareness that frontal lobe changes, which mediate cognitive function, are equally important to identify and consider, to provide optimal care (Murphy et al., 2016; Phukan et al., 2007)

Fronto-temporal decline in MND occurs across a spectrum (Oliver & Turner, 2010; Phukan et al., 2007). An estimated 15% of pwMND develop a frank fronto-temporal dementia

(Phukan et al., 2007), or Amyotrophic Lateral Sclerosis Fronto-temporal dementia (ALS-FTD). Altered eating patterns such as compulsiveness and increased bolus sizes are recognised in FTD, both with/without the presence of MND (Ahmed et al., 2016; Langmore et al., 2007). However, approximately 35% of pwMND experience mild-moderate cognitive impairment (Caga et al., 2019) (not meeting the criteria for FTD diagnosis), and it is unclear how these changes interact with oropharyngeal swallowing function.

Mild-moderate cognitive impairments may have less overt symptoms or have a range of characteristics that are atypical of FTD. In addition, impaired motor function and/or dysarthria associated with MND may mask these more subtle changes (Caga et al., 2019). Mild–moderate cognitive impairment in MND may manifest as apathy, reduced insight (Flaherty-Craig et al., 2011), impaired social cognition, frustration intolerance or irritability (Murphy et al., 2016) or mental rigidity (Phukan et al., 2007). These changes impact on a pwMND's decision-making capacity (Khin et al., 2015) and adaptability. For example, non-invasive ventilation (NIV) is routinely used in MND to manage sleep disordered breathing and dyspnoea, and is linked to prolonging survival (Martin et al., 2014). In addition, gastrostomy is considered prophylactically and/or when swallowing function is not safe to sustain adequate nutrition or hydration orally, or when hypermetabolism associated with MND causes weight loss and malnutrition (Bouteloup et al., 2009). However, pwMND with cognitive impairment may be more likely to refuse these interventions (Martin et al., 2014). It is not known if mild-moderate cognitive impairment also has implications for oropharyngeal swallowing management, including implementation of a modified diet or safe swallowing strategies.

Research has explored swallowing function in MND however, little is known about how the experience, assessment and management of oropharyngeal swallowing may be impacted by cognitive impairment. This is important as oropharyngeal swallowing assessment often involves patient self-report, and interventions include dietary restriction and/or modification

and strategies, such as postural adaptation (Rumbach et al., 2018). These require a pwMND to actively participate and enact, and consequently cognitive engagement is required (Egan et al., 2020). Further, there is evidence that FTD is associated with changes to eating behaviour and food preference (Langmore et al., 2007). However, little is known about the impact of mild-moderate cognitive changes, not meeting FTD criteria, on oropharyngeal swallowing function in pwMND. In addition, how mild-moderate cognitive impairment affects pwMND's capacity to make decisions regarding their care, provide consent to swallowing interventions, and implement management plans is not understood. This is important as swallowing problems are associated with poorer outcomes in MND, and the added presence of cognitive impairment is likely to further complicate swallowing management and outcomes (Paris et al., 2013).

The impact of mild-moderate cognitive impairment associated with MND is less prominent in the literature than research about MND-FTD. Therefore, our intention was to retain a narrow search scope that focussed on oropharyngeal swallowing function, in individuals presenting with mild-moderate cognitive impairment associated with MND. We conducted a primary systematic scoping review to synthesise research evidence about how pwMND with associated mild-moderate cognitive impairment enact commonly implemented approaches, which modify oropharyngeal swallowing function, such as swallowing compensatory strategies and diet modifications, as these are often implemented by speech pathologists or other allied health professionals. As such, this review sought to answer the research question: *How does cognitive impairment impact on a pwMND's ability to understand and manage oropharyngeal swallowing function?*

To expand our understanding, we conducted a secondary search with broadened search terms encompassing cognition (e.g., FTD) and dysphagia (e.g., eating & mealtimes) to investigate the parameters of evidence.

Method

This scoping review applied the methodological framework of (Arksey & O'Malley, 2005), to guide the development of the research question and search strategy, identify relevant studies, chart and analyse the data and report the results. Subject headings and key words (MND + cognition + dysphagia) as shown in Table 3 were searched.

Table 3.

Scoping review search terms related to MND and cognition and dysphagia

Motor neurone disease		Cognit*		Dysphagia
Motor neurone disease		Behaviour		Dysphag*
MND		Awareness		Swallowing
Amyotrophic lateral		Perception		disorder
sclerosis		Self-perception		Swallowing
ALS		Insight		impair*
Progressive Muscular		Symptom		Deglutition
Atrophy		Symptom Assessment		Deglut*
PMA	AND	Knowledge	AND	Deglutition
Progressive Bulbar		Opinion		disorders
Palsy		Attitude		Deglut* disorders
PBP		Complaint		*Eat**
Primary Lateral		Pre-diagnosis		*Meal**
Sclerosis		Early diagnosis		*Feed**
PLS		Memory		*Appetite*
Kennedy's disease		*Frontotemporal dementia*		*Mastication*
Neurodegenerative		*Executive Function*		*Drinking*
disease		*Neurocognitive disorders*		
		Dementia		

Note: Italics denotes search terms added in secondary search. MND: Motor Neurone Disease

Inclusion/exclusion criteria

This scoping review intended to explore how mild-moderate cognitive decline in MND impacts oropharyngeal swallowing function and management. Therefore, articles that included an MND population and reported on an outcome measure relating to both cognition AND swallowing were included. Features related to swallowing included the diagnosis and/or management of oropharyngeal dysphagia, or adherence to safe swallowing strategies. Inclusion required objective measures or direct clinical observation of swallowing. Articles were excluded if no objective measures or direct clinical observation of swallowing were reported on, the population was not MND, not human, not adult, or not available in English.

Documentation and evaluation of search

The primary search (unrestricted dates) was executed across: MEDLINE, SCOPUS, CINAHL, PsychINFO, Emcare and Google Scholar, in May 2019 and repeated in November 2019 to include newly published articles. Articles employing various designs and data collection methods, along with information derived from the grey literature sourced via Google searches were included. The reference lists of the reviewed full-text articles were also searched.

Relevance Screening

Two reviewers conducted independent screening. A third reviewer was consulted if agreement could not be reached. Using Covidence © (see Figure 15) duplicates were removed, and title and abstract screening was conducted on 683 articles. From these, 636 articles were excluded, and 47 full-text articles from allied health and medical journals, MND clinical guidelines and case studies were retrieved and assessed for eligibility.

Using the inclusion criteria, the full texts of the 47 articles were independently reviewed by two reviewers. A third reviewer was consulted in case of disagreement. None of the full text articles reviewed directly addressed the research question for the following reasons: 39 articles did not directly meet the inclusion criteria of cognitive impairment in MND in relation

to swallowing, three were not available in English, two were duplicates, two reported on a paediatric population, and one did not report on MND (Figure 15).

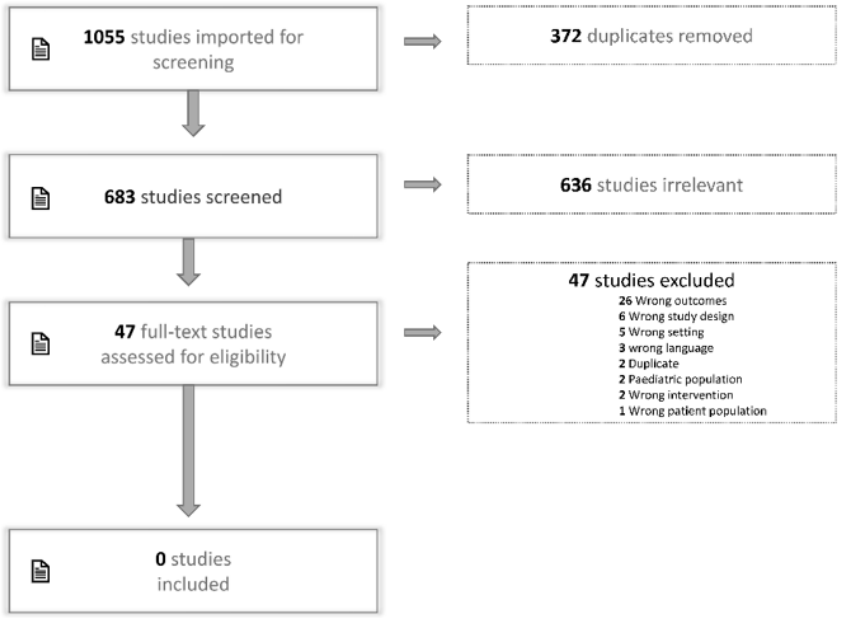

Figure 15: PRISMA 1 - Primary literature search (*PRISMA: Preferred Reporting Items for Systematic reviews and Meta-Analyses*). Image created by thesis author.

As the first screening did not yield articles relevant to the research question, the research team, in consultation with a medical librarian, devised a secondary search strategy to include broader terms encompassing dysphagia (e.g., eating, feeding) and cognition (e.g., Fronto-temporal dementia) as shown in italics in Table 3. The secondary search was executed across Medline, Psych Info and Emcare in July 2020, and the dataset imported into Covidence © for screening and review (Figure 16). Duplicates were removed, and title and abstract screening was conducted on a further 762 articles. From these, 753 articles were excluded, and 9 full-text articles were retrieved and assessed for eligibility.

Using the inclusion criteria, the full text of the 9 articles were critically reviewed by two reviewers independently who agreed that the additional full text articles did not address the research question for the following reasons: 8 articles did not directly meet the inclusion

criteria of cognitive impairment in MND and dysphagia, and one was not available in English (Figure 16).

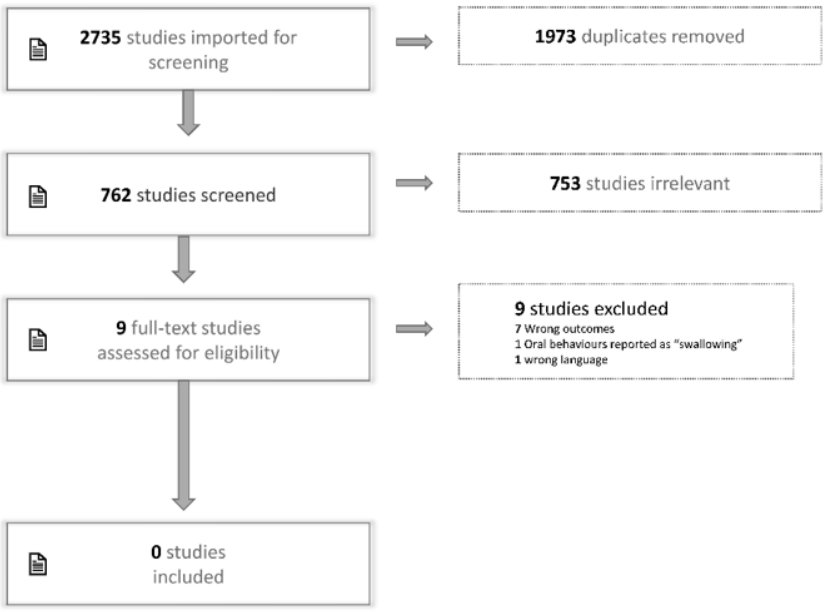

Figure 16: PRISMA 2 - Secondary literature search.
(*PRISMA: Preferred Reporting Items for Systematic reviews and Meta-Analyses*). Image created by thesis author.

While no eligible studies were found, empty reviews provide an important opportunity to gain an understanding of the breadth of current research evidence, identify gaps in knowledge and inform future research direction (Lang et al., 2007). Consequently, we conducted a thematic analysis, guided by the Braun and Clarke framework (Braun & Clarke, 2006) to identify themes within the 56 articles that underwent full text review as these formed a body of literature that referenced swallowing or dysphagia AND cognition in MND. Before undertaking the thematic analysis, we removed 27 articles that did not meet the original inclusion criteria. Whilst these included a reference to swallowing and cognition, their content was not relevant to the research question (i.e., pharmacology, genetics, not specific to MND). Therefore, 29 articles were included for thematic analysis. Please refer to Appendix 3 for Supplementary Table I for a complete list of the reviewed articles.

To extract these data, the researcher re-read the full text of each included article and made a direct copy of any text which referenced swallowing or dysphagia AND cognition in MND to a Microsoft excel spreadsheet. The lead author inductively coded data from the 29 articles into NVivo Pro 12. This data extraction was then analysed by another member of the research team. The research team reviewed the coded categories, then agreed on subcategories. Once categorisation was complete, *thematic analysis was conducted on the extracted data, whereby* patterns within the data were identified, discussed and a consensus was reached on key themes. Please refer to Supplementary Table II (Appendix 4) for the open coding and categorisations of article excerpt data and the relationship to main theme.

Findings

The thematic analysis of literature that included terms related to dysphagia and cognitive impairment in MND identified three key themes. These themes did not specifically address the research question, which verified the negative result of the initial scoping review. The links between the main three themes identified in the thematic analysis and the importance of understanding the impact of cognitive impairment when managing oropharyngeal swallowing function in MND will be examined in the discussion.

Theme 1: Early and regular specialised multidisciplinary management of MND achieves better outcomes

Recommendations for specialised multidisciplinary clinics to manage MND featured prominently within the literature that referred to both cognition and oropharyngeal swallowing function (Orrell, 2016; Rodriguez de Rivera et al., 2011; Tiryaki & Horak, 2014). Early identification, timely interventions and a proactive approach to symptoms were recommended. This alleviated patient distress, caregiver burden and reduced medical emergencies from falls or respiratory distress, which may result in hospital admission. Review appointments were suggested every three to four months depending on rate of decline.

> The complexity and progressive nature of MND often necessitates multidisciplinary care, which leads to better outcomes and less utilisation of emergency care (Tiryaki & Horak, 2014).
>
> MND care is best delivered by a multidisciplinary team. Most patients do best when they are seen every 3–4 months; however, visits should be scheduled based on rate of disease progression (Oliveira & Pereira, 2009).

Outcomes reported in the included literature suggest that a specialised clinic consists of (but is not limited to) a neurologist, specialist nurse, dietitian, physiotherapist, occupational therapist, respiratory physiologist, speech-language pathologist, and palliative care doctor to optimise care (Orrell, 2016). Compared to management within a general neurology department, multidisciplinary management of MND was reported to extend survival by approximately seven months.

> …compared 344 patients in MDC to patients in general neurology care (GNC) and found 7.5-month longer survival in the MDC cohort (Williams et al., 2014).

This outcome was attributed to earlier access to specialised therapies, a coordinated approach to care, and improved capacity for HCPs to anticipate rates of decline associated with phenotype. Regular monitoring facilitated a more proactive approach to treatment, and the detection of sub-clinical symptoms such as a decline in cognitive function.

> …treatment at multidisciplinary units does not seem to alter the neurological course of the disease but rather it favours the application of respiratory and nutritional care, as well as the detection and treatment of symptoms of depression and cognitive impairment (Rodriguez de Rivera et al., 2011).
>
> We believe that the high levels of patients with symptoms of depression and cognitive impairment are due to the fact that the patients are monitored at a

> multidisciplinary unit and their follow-up by different specialists according to the protocol might make it easier to detect sub-clinical findings (Rodriguez de Rivera et al., 2011).

Despite agreement within the reviewed literature that better outcomes result from a multidisciplinary care approach, multidisciplinary management of oropharyngeal swallowing function, in the presence of cognitive decline was not explicitly discussed in the included articles.

Theme 2: Cognitive decline impacts on management and survival time

The included articles reported that cognitive impairment occurs in up to 50% of pwMND, is more common in bulbar onset, and predicts worse prognosis and shorter survival, but the included articles did not include a direct link with oropharyngeal swallowing function.

> Cognitive problems are important, as they confer a worse prognosis (Elamin et al., 2013; Jenkins et al., 2014).

> …such dysfunction is associated with poorer prognosis (Eisen et al., 2014).

In MND, cognitive impairment negatively impacts on decision-making, insight, adherence and treatment uptake (Oliveira & Pereira, 2009), and the efficacy of some treatments may be reduced if instructions are not able to be followed**.** Cognitive impairment is also reported to cause apathy, irritability and alterations in behaviour.

> Cognitive impairment may decrease acceptance of care interventions and impair decision making (Tiryaki & Horak, 2014).

> Shortened survival and a higher rate of non-compliance with recommendations for non-invasive ventilation and feeding tubes are seen in patients with MND with frontotemporal dysfunction (Vengoechea et al., 2013).

> MND is associated with a frontotemporal type of dementia, and it is associated with a negative impact on survival. Cognitive impairment has been demonstrated in 20–50% of patients with MND (Williams et al., 2014).

Cognitive impairment may also impact on the provision of management and care for pwMND. Carers and HCPs may mistake behaviour changes related to cognitive impairment as rudeness, argumentativeness or stubbornness (Tiryaki & Horak, 2014). Therefore, pwMND with associated cognitive impairment are likely to have different care needs to manage swallowing impairments, compared to those with intact fronto-temporal function or those with a diagnosis of MND-FTD.

> Tailor all discussions to the person's needs, taking into account their communication ability, cognitive status and mental capacity (NICE, 2016).

In parallel with the need for regular multidisciplinary input from Theme 1, the literature reviewed identified that pwMND benefit from early cognitive screening with regular assessment and monitoring for change, but direct implications for swallowing management could not be inferred from the included literature.

> Early detection and regular assessment of cognitive deficits can improve care by making the patient more proactive and leading to earlier end-of-life discussion (Maiser & Tiryaki, 2017).

Theme 3: Oropharyngeal swallowing impairment occurs in nearly all pwMND and is a serious symptom of the disease

In the reviewed literature, that included concepts of both cognition and oropharyngeal swallowing function, swallowing impairment was described as a serious symptom of MND, increasing the risk of choking, aspiration and weight loss, which is associated with higher rates of malnutrition and dehydration. Most of the articles linked worsening dysphagia

symptoms with disease progression (Burgos et al., 2018; Oliveira & Pereira, 2009; Onesti et al., 2017; H. Stavroulakis et al., 2014).

> As dysphagia progressed, most participants described how eating and drinking became increasingly difficult (H. Stavroulakis et al., 2014).

> …most MND patients experience some degree of swallowing difficulty at some time during the course of the disease, with progressive worsening. Drooling, dehydration, malnutrition with weight loss and aspiration are all associated with dysphagia (Oliveira & Pereira, 2009).

Similar to elements of Theme 1, prompt assessment and ongoing monitoring of swallowing function every three months was recommended for all pwMND. Examination of the appearance and movement of the tongue, lips, and jaw, combined with self-reported swallowing questionnaires were frequently reported assessments. The recommended instrumental assessments for swallowing function in MND were Videofluoroscopic Swallowing Study (VFSS) and Flexible Endoscopic Evaluation of Swallowing (FEES).

> With videofluoroscopic evaluation of swallowing, 75% of patients had dysphagia (Burgos et al., 2018).

The recommendation for enteral feeding featured prominently in the reviewed literature, with the insertion of Percutaneous Endoscopic Gastrostomy (PEG) recommended when the decline in oropharyngeal swallowing function makes oral feeding unsafe or impossible. PEG was also recommended as a proactive intervention to maintain a high body mass index and avoid weight loss associated with hypermetabolism.

A gastrostomy tube should be discussed with individuals with significant dysphagia who continue to lose weight despite their best efforts (Golaszewski, 2007).

Biomechanical and functional characteristics of swallowing in MND were discussed in the

included literature (Burgos et al., 2018; H. Stavroulakis et al., 2014; Williams et al., 2014). Oropharyngeal swallowing impairments were described as impaired chewing, reduced tongue strength, inadequate bolus control and propulsion, reduced pharyngeal constriction and/or impaired bolus passage across the upper oesophageal sphincter. Aspiration was prominently reported as a risk associated with all dysphagia symptoms. Patients self-reported dysphagia symptoms to include coughing, choking, and longer, more difficult mealtimes.

They reported that coughing and choking on attempting to swallow became more common and meals were also becoming difficult, prolonged, tiring and stressful (H. Stavroulakis et al., 2014).

Whilst cognitive impairment associated with MND was discussed in each of the included studies that also described oropharyngeal swallowing function, it was not the research focus of any of the included studies. Rather, pwMND with cognitive impairment were consistently excluded from the reviewed studies that also included swallowing.

Discussion

The purpose of this scoping review was to investigate how mild-moderate cognitive impairment associated with MND impacts on a pwMND's ability to understand and manage oropharyngeal swallowing function. The findings of our primary search identified that this relationship has not yet been explicitly investigated. This may be because the role of cognition in MND has only recently received attention and the interplay between mild-moderate cognitive impairment and other symptoms of MND has not yet been fully investigated.

Our secondary search included expanded search terms that were relevant to our research question to explore the intersection between oropharyngeal swallowing and cognitive impairment in MND more broadly. Of note, one study included in the full text review of the

secondary search (Ahmed, et al., 2016) investigated cognition and eating behaviour in MND, MND plus mild-moderate cognitive change, MND-FTD and FTD. On further investigation of the methods, we determined that this study did not meet inclusion criteria, as information on swallowing function was not based on direct clinical observation or objective assessment, but on carer report. Whilst this study used subjective measures to describe swallowing behaviours, it does provide critical preliminary information that directly links changes to swallowing and eating behaviours with cognitive changes for pwMND. As such, it is important to further evaluate how the spectrum of cognitive change in MND impacts on how pwMND understand and manage an oropharyngeal swallowing impairment.

Whilst the thematic analysis showed that cognitive impairment and swallowing impairment are separately perceived as important symptoms of MND, it also verified that research has not directly explored a relationship between cognitive function and oropharyngeal swallowing function in this population. Whilst the three identified themes did not directly answer our research question, they assist in situating the identified research gap into the context of how cognitive impairment may affect the assessment, management, and care of pwMND and their carers more broadly.

The first key theme verified existing research, demonstrating that better health outcomes are achieved if the management of MND is delivered by a specialised MDT within a patient-centred care framework (Corcia & Meininger, 2008; Fullam et al., 2016). However, there was no literature that connected this finding directly with oropharyngeal swallowing. In addition, patient-centred care relies on two-way communication to create a mutually agreed management plan (Paynter et al., 2019). As cognitive impairment may impact on the information exchange between HCPs and patients, the impact of this for how oropharyngeal swallowing function is managed as part of a patient-centred care framework requires more exploration. Specifically, cognitive impairment has been shown to be a barrier to the decision-making processes (Hogden et al., 2013). For example, cognitive impairment in

MND may manifest as a lack of insight, inflexibility, or mental rigidity, and consequently, a pwMND may not understand the symptoms of the disease or the need for management. Similarly, cognitive impairment may also impede relatively complex decision-making regarding swallowing management, including mealtime adaptations and enteral feeding. This has the potential to jeopardise swallowing safety of pwMND as well as compound any concerns and anxieties felt by caregivers in relation to mealtimes.

Providing early education to pwMND and their carers about cognitive impairment that can accompany MND may assist in the recognition of changes, alleviate facets of care-giver burden, and aid in proactive decision-making and consent. Current MND clinical guidelines (National Institute for Health and Care Excellence, 2016) recommend that specialised MND clinics include cognitive screening both at initial assessment, and as regular monitoring to ensure any decline is quickly identified and can then be considered in the management plan. This was also a prominent theme in the analysed literature, however direct implications for oropharyngeal swallowing management could not be inferred.

The second theme described how cognitive impairment impacts on MND management. For example, pwMND who also have cognitive impairment have been shown to be less likely to opt for life prolonging medical interventions and may have greater difficulty using and tolerating equipment (Martin et al., 2014). It is currently not known if cognitive impairment also impedes a person's ability to understand complex alterations to their diet and eating or impacts on their acceptance and uptake of oropharyngeal swallowing management strategies. As outlined in this second theme, the emerging literature on the role of cognition in MND highlights the importance of considering cognitive function and capacity when planning and implementing ongoing care and swallowing interventions.

The third key theme identified that oropharyngeal swallowing management is critical in MND to reduce respiratory complications and to ensure adequate caloric intake. However, studies investigating swallowing in MND typically exclude participants with evidence of cognitive

impairment (Ertekin et al., 2000; Fattori et al., 2017; Onesti et al., 2017; Ruoppolo et al., 2013; Tabor et al., 2016). This appears to be a gap in the current literature as most pwMND will develop an oropharyngeal swallowing impairment as the disease progresses (Onesti et al., 2017) and up to 50% of pwMND also develop a cognitive impairment (Watermeyer et al., 2015). The findings of studies excluding pwMND with cognitive impairment lack validity for half of the MND population. Additionally, the results of studies which did not control for the presence of cognitive impairment must be interpreted in the context that the interaction between swallowing and cognitive impairment, is unknown. As such, cognitive function is an important factor to measure and control for when investigating swallowing in MND, particularly when typical assessment methods that include self-reporting rely on insight.

Self-reporting is an important tool in dysphagia assessment. However, it is critical that the self-reported information is triangulated with information relating to individual's cognitive status. Self-assessment tools such as the EAT-10 (Belafsky et al., 2008) and the Sydney Swallow Questionnaire (Szczesniak et al., 2014) are routinely used in clinical care of pwMND to determine a person's level of swallowing function and to guide HCPs' management planning. However, future research is needed to explore if and how cognitive impairment affects the validity of the outcomes of these tools when used in MND assessment (Plowman et al., 2016).

As reported in the reviewed literature, instrumental assessment of swallowing in pwMND should be conducted via VFSS or FEES. However, these assessments rely on the person being able to follow directions, swallow on cue and for FEES, tolerate invasive scoping. As such, cognition screening is recommended prior to instrumental assessment in order to determine suitability (Palmer et al., 1993), as cognitive difficulties have been identified as a contraindication for conducting these instrumental examinations. Therefore, an understanding of cognitive ability in a pwMND is vital prior to conducting an instrumental swallowing assessment (Martin-Harris & Jones, 2008).

Clinical management of impaired swallowing often combines diet modification, compensatory measures, diet restrictions and postural adaptations to ensure safe oral intake (Ruoppolo et al., 2013). It is not currently known if cognitive impairment impacts on pwMND's ability to understand their swallowing problem, follow recommendations, or enact strategies. Further, it is not yet known if decline in swallowing motor function occurs in parallel with frontal lobe deterioration, or if the characteristics of swallowing function are altered in the presence of cognitive impairment. As it has been shown that bulbar onset MND increases the risk of developing both impaired swallowing and cognitive impairment, investigating the interaction of these two variables may provide valuable insights into the disease.

The identified themes revealed that regular and ongoing MDT management, which includes cognitive assessment, are commonly reported for MND management. However, in relation to swallowing management, this review suggests that cognitive assessment and consideration of the impact of cognitive changes on swallowing management, have been omitted. In light of the research question, and particularly our focus on pwMND with associated mild-moderate cognitive difficulties, these themes reflect and raise awareness of, the complexity in the intersection between the holistic, person-centred, team-based approach identified across the themes, and challenges associated with informed decision making and risk awareness related to swallowing in relation to a person's cognitive skills. These prominent themes in the MND literature suggest that more research is needed to understand how mild-moderate cognitive impairment influences how a person-centred, team-based approach to swallowing management can be enacted.

Limitation

The narrow focus of this review was an intended strategy to explicitly investigate the variables of mild-moderate cognitive impairment and oropharyngeal swallowing function. To explore articles that were the most relevant to the research question, the number of elements in the search strategy were minimised to optimise the results (Bramer et al., 2018). The studies analysed in this review did not explicitly answer our research question and we are unable to draw specific conclusions. The thematic analysis conducted was limited to the literature which contained references to both dysphagia and cognition but were not necessarily informative of both. In addition, due the spectrum of cognitive change that occurs in MND, it is difficult to resolve terms that differentiate between mild-moderate cognition and the criteria required for an FTD diagnosis.

Future research

The findings of this study demonstrate that research which investigates how cognitive impairment influences oropharyngeal swallowing function and management in pwMND is lacking. Future studies could examine this relationship, and further research may also explore the incorporation of MND specific cognition screening tools to detect impairment early in the disease course. Inclusion of participants with cognitive impairment will ensure an accurate representation of the MND population. Whilst there has been preliminary descriptive exploration of swallowing function in pwMND with associated cognitive impairment (Ahmed et al., 2016), observations in this study were captured via carer report, foregrounding the necessity for robust, objective measures of swallowing function for this population. Further, studies which aim to identify if swallowing characteristics differ between those with/without cognitive impairment may help to inform the assessment and management of swallowing interventions.

Conclusion

The impact of mild-moderate cognitive impairment associated with MND on oropharyngeal swallowing function is unknown. The primary analysis of this review yielded no results that specifically addressed the interaction between mild-moderate cognitive impairment associated with MND and oropharyngeal swallowing function. Subsequent thematic analysis identified three prominent themes in the MND literature that contained information about both oropharyngeal swallowing function and cognition.

Cognitive impairment may have implications for decision-making, treatment uptake and adherence to swallowing management, potentially increasing the risk of unsafe swallowing and increasing caregiver burden. In line with the National Institute for Health and Care Excellence (2016), we suggest that cognitive screening should form part of routine MDT MND management, early in diagnosis and repeated as part of regular disease monitoring. Communication approaches can then be adjusted to enable best patient-centred care. A proactive approach to cognitive impairment may not only benefit the pwMND but may also relieve some of the burden felt by carers.

The previous study identified that to date, there have been no investigations into the interaction between cognitive and behavioural changes in MND and dysphagia. As cognitive and behavioural changes have been shown to impact on decision-making and adherence to other MND management strategies (Greenaway et al., 2015; Oliver et al., 2011), it is important to evaluate cognitive and behavioural changes in the context of swallowing function. Therefore, to better understand pwMND and carers' perspectives and to develop a deeper and richer understanding of swallowing management in MND in the home context, we designed a longitudinal ethnographic study. In this qualitative study, I originally planned to conduct observations of the family mealtime with pwMND and their carers. An ethnographical approach was chosen, as it allows researchers to immerse themselves in the environment they wish to study, enabling sustained observations and providing opportunities to develop rich insights. Ethics for this research study was approved in February 2021 (Appendix 2)

Originally, the ethnographic study was planned to take place from May 2021 to March 2023, with several re-attempts to recruit participants during this time. However, this period coincided with the Covid-19 pandemic. During the pandemic, the risks for pwMND to participate in research was deemed too great by many, as pwMND already have compromised respiratory function. Being exposed to the Covid-19 virus by external people entering the family home was deemed an unnecessary risk. The world was experiencing great uncertainty during this time, and as no participants were recruited over a long period, it became evident that this research study was no longer feasible. Consequently, we decided to cease recruitment in March 2022. We notified the relevant human research ethics committee and pivoted the study towards interviewing pwMND remotely. In the following chapter, I present the study background and methods of the planned ethnographic study as intended, in order to inform the reader of the intended approach and to reflect the effort that was invested as part of this program of research.

CHAPTER 4: HOW DO PEOPLE LIVING WITH MND UNDERSTAND AND MANAGE A SWALLOWING DISORDER? AN ETHNOGRAPHIC STUDY.

Introduction

Almost all pwMND develop dysphagia over the course of disease progression. (Burgos et al., 2018; Murono et al., 2015; Onesti et al., 2017). Speech pathologists are an integral part of an MND MDT, assessing, diagnosing and implementing dysphagia management strategies. These strategies may consist of diet modifications, or behavioural adaptations. For example, a common dysphagia recommendation to support a person to eat and drink safely is to recommend a modified bolus size (American Speech-Language-Hearing Association, 2024), e.g., by reducing the amount of food and fluid a person takes in each mouthful. However, this seemingly simple strategy requires cognitive engagement to understand and enact consistently when eating and drinking. One must first understand the swallowing impairment and change of function, and then appropriately and independently evaluate the mouthful size to sustain safe eating and drinking.

Similarly, a more permanent management approach for severe dysphagia, enteral feeding, requires a pwMND to understand the functional impairment, understand the procedure and provide informed consent to the procedure. After implementation of gastrostomy, a pwMND then must adhere to recommendations of use and daily maintenance to ensure acceptable levels of hygiene and proper functioning. However, in parallel to dysphagia, up to 50% of pwMND also experience cognitive and behavioural changes (Strong et al., 2017). These changes have been shown to impact on a pwMND's ability to understand new and complex information (MND Australia, 2020a; Paynter et al., 2019). In addition, a pwMND may have impaired reasoning, impaired problem solving, reduced insight, reduced awareness, and difficulties with planning or organising (MND Australia, 2020a). However, the interactions of cognitive and behavioural changes in MND and dysphagia have not yet been explicitly

investigated (Francis et al., 2021) (Chapter 3). Cognitive and behavioural changes have previously been linked to a reduced capacity in pwMND in relation to decision-making, treatment uptake and treatment adherence (Chiò et al., 2012; N. H. Martin et al., 2014). For example, non-invasive ventilation (NIV) is routinely used in MND to manage sleep disordered breathing and dyspnoea, with this intervention being linked to prolonged survival (Martin et al., 2014). However, pwMND who also have cognitive impairment, are more likely to refuse this ventilatory support (Martin et al., 2014). In addition, pwMND who also have cognitive and behavioural changes have greater difficulty using and tolerating NIV equipment (Oliver, 2019).

Consequently, studies which explore the relationship between cognitive and behavioural changes associated with MND and dysphagia are needed. These investigations are necessary to determine if and how cognitive and behavioural changes affect the way people experience swallowing. Specifically, it is of interest to evaluate how pwMND, their carers and families with or without cognitive and behavioural changes make informed decisions on dysphagia management, and how they implement safe swallowing strategies in the home environment, whilst maintaining adequate nutrition and hydration. A better understanding of these factors to inform clinical practice and improve patient care.

Therefore, the aim of this research was to develop an understanding of how pwMND who may or may not have a cognitive and behavioural changes, understand and manage dysphagia within their home environment, and how safe swallowing management plans are enacted. To allow for this exploration, a longitudinal ethnographic study was planned as this would allow for the development of a deeper understanding of how speech pathology management strategies are received by families experiencing MND, through long-term participant observation. The following research questions were to be addressed:

4. What are the experiences of people living with MND, their carers and family members of understanding and managing dysphagia?

5. What are the facilitators and barriers for people living with MND for implementing and/or adhering to a swallowing management plan?
6. How do cognitive and behavioural changes impact on the understanding and management of swallowing impairments for pwMND?

Proposed method

Our aim was to employ a descriptive longitudinal ethnographic approach with in-home observations, as a means of data collection. Mealtimes within the home (or social outings) as well as family interactions were the focus of this research project. An ethnographical approach was chosen, as there is limited literature available that investigates the lived experience of dysphagia and MND, and consequently, a qualitative research methodology was required to explore this unique phenomenon. Ethnographical studies provide an emic view and give researchers insight into people's behaviours, experiences and attitudes. Therefore, observations and unstructured interviews were planned to be the central method for data collection. As shown in Appendix 2, ethics approval for this project was gained from the HREC at Flinders University (HREC ID: 2850) in 2021.

Proposed recruitment

We aimed to recruit approximately ten pwMND and ten primary carers. Any person with a confirmed or probable diagnosis of any phenotype of MND, and their carer would be eligible to participate. PwMND who were receiving palliative care support would not be excluded, as palliative care may commence at the time of MND diagnosis. However, any person with a diagnosis of MND-FTD, pwMND who resided in full time care, or who live alone, would be excluded, because of the additional burden participating in research may cause. Additionally, as this study was investigating how dysphagia was managed behaviourally in the home environment, pwMND who were nil by mouth, had no functional swallowing and/or received all nutrition and hydration via enteral feeding, were also excluded from participation.

Participant recruitment took place via social media posts and a MND SA newsletter mail out. To follow are examples of social media posts and newsletter recruitment (Figure 17)

Instagram, Facebook and X (formerly Twitter),

@Flinders PhD Researcher and speech pathologist, Rebecca Francis is conducting a study which is investigating #swallowing in #Motor neurone disease. This study will investigate the lived experiences of managing a swallowing problem (#dysphagia) in #MND. If you would like to receive more information, please contact Rebecca on 08 82012811 or *rebecca.francis@flinders.edu.au* @bekfrancis @swallowneurolab @CFI #slpeeps

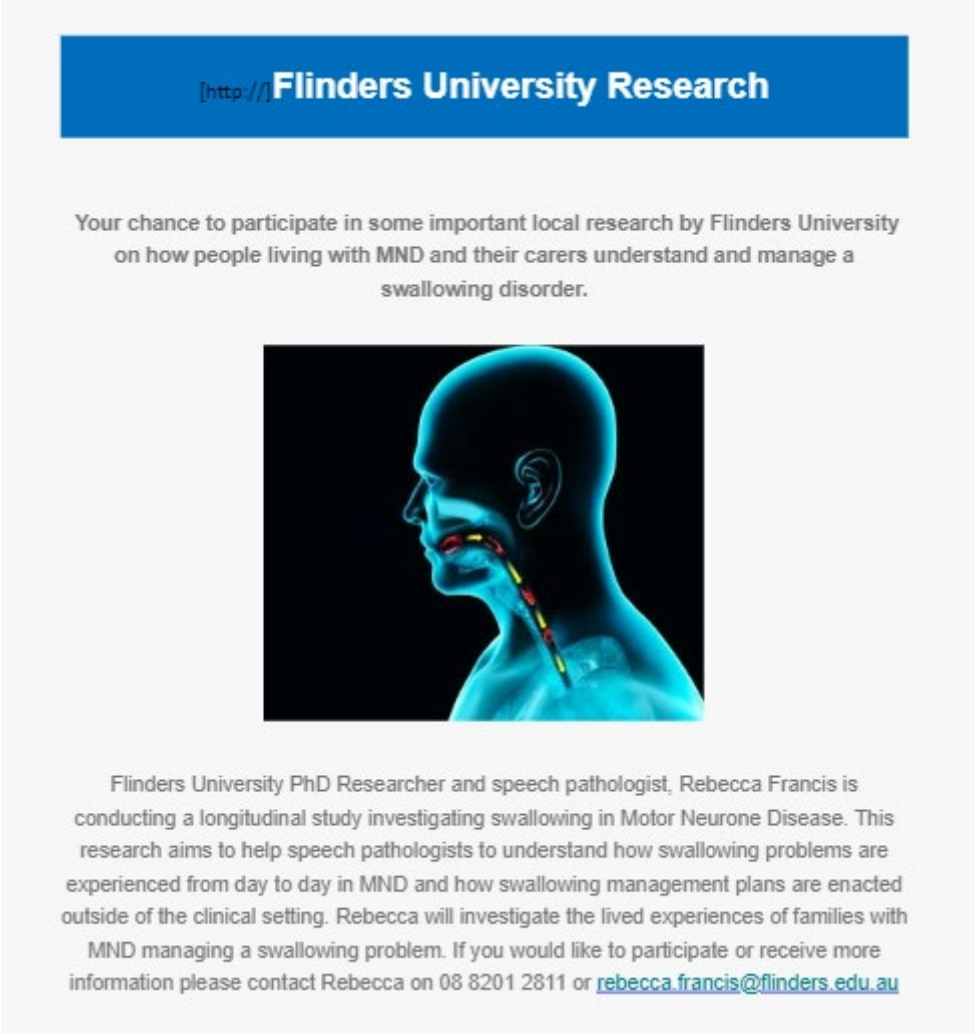

Figure 17: Example of the recruitment notice included in the MND SA newsletter, emailed to MND SA participants on the 16th of June 2021.
(MND: Motor Neurone Disease; SA: South Australia)

Proposed data collection

Frequency of home visits

The current MND guidelines suggest that both swallowing function assessment and cognitive screening should take place at regular intervals of three months over the course of the disease (Burgos et al., 2018; National Institute for Health and Care Excellence, 2016). As a longitudinal study, it was planned to observe consented participants within their home environment, at approximately three-monthly intervals. As this study explored dysphagia over the course of MND, it was also planned that participants would be involved with routine observations and unstructured interviews over a 1-2-year period.

Proposed cognition and behaviour screening

In order to monitor for any changes to cognition or behaviour, the Amyotrophic Lateral Sclerosis Cognitive and Behavioural Screen (ALS-CBS) (Woolley et al., 2010) was to be conducted with pwMND (cognitive subtest) and their primary carer (behavioural subtest) at each visit (not more than three monthly). As a previously validated screening tool, the ALS-CBS identifies frontotemporal spectrum dysfunctions in pwMND and continues to be suitable for screening as motor function deteriorates (Woolley et al, 2010). The ALS-CBS was to be conducted to record any changes to cognition and/or behaviour over the course of the disease, and the results from each screen were to be forwarded, with consent, to the participant's medical team.

It had been planned to attend the participants' home (or mutually agreed meeting place) during family mealtimes with each visit to take a maximum of 3 hours. This planned duration allowed for data collection to occur via both silent observation and unstructured interviews whilst families undertook meal preparation and ate their meal.

The first 10 minutes of each visit was allocated to conduct a general conversation with the family and to discuss any questions or concerns. Following this, the ALS-CBS was to be completed with the pwMND, whilst the primary carer completed the behavioural section. Then, an observational period within the home was planned, to collect data on how meals were prepared and of the mealtime experiences of families. The planned observation was not directed, and the intent was that the visits would have a minimally intrusive presence on the family to allow for observation of the family dynamics and interactions. During the observations, field notes would be recorded about the mealtime experience for both the pwMND and the carer. Specifically, the ease or difficulties that may occur whilst both preparing and eating a meal, as well as the ease or difficulties that may occur related to the dysphagia management plan. Further, field notes would be recorded to capture interactions between the pwMND and their carer.

At the end of the observational period, anticipated to correspond with the completion of the meal, it had been planned to conduct a debriefing session with the family that would explore questions designed to help address the research questions. The aim was to develop a richer understanding of the thoughts and feelings of both the pwMND and the carer. Further, to ensure accurate representation, it was planned to audio record the conversations, with consent, to create transcripts.

No participants were recruited for this study, and subsequently the project was stopped in March 2022.

Several attempts were made to recruit participants between April 2021 and March 2022 before a collective decision was made by the research team to close the study. In August of 2021, one family requested information via email. Subsequently, they were provided with the PICF via return email. However, this family advised shortly after that they had decided not to proceed. Following this, the study details were reposted on the social media platforms as mentioned above. Further, direct emails were sent to colleagues working with families with

MND with the PICF attached, to ask them to provide information of the study to people who met the inclusion criteria. In addition, MND SA was approached to enquire if they had received any feedback from families with MND on the study. During this discussion, the researcher proposed creating a short video for inclusion in the MND SA newsletter and/or attending an MND SA information sessions to provide information of the research project in this space. However, these suggestions were not agreed to.

In October of 2021, I was advised by a speech pathologist from an MND clinic that a family had requested information about the research project, but unfortunately this family did not contact me.

It is not entirely clear why there was no uptake for this study, however at the time of recruitment, the global pandemic was at its peak, and all people with an already compromised immune or respiratory system were at increased risk of serious complications or death from the virus. It is understandable that pwMND would not want to participate in any activities that could increase their exposure to Covid-19. This view was echoed from the Client Services Manager at MND SA, who advised that attendance at their 'You, Me and MND' sessions was also reduced. Another possible explanation for lack of participation was potentially related to the time commitment of this study. It may have been viewed as too intensive, given that this was a longitudinal study, with regular visits planned over an extended period of time.

As a result, the decision was made to pivot this program of research and instead to explore research options that enabled online contact with families

As I was not able to recruit participants for the planned ethnographic study, potentially due to the impact of Covid-19, it became evident that a pivot was needed (as shown in Figure 1). A change in research direction, which was designed around online recruitment, would enable my PhD research to continue during a global pandemic. Subsequently, this program of research was reviewed by the research team and revised. The new approach continued to align with the overall aim of this research; to investigate the interaction between cognitive and behavioural changes and dysphagia in MND. However, a shift to an online focus allowed for data collection to continue, but not be reliant on face-to-face contact with families living with MND. At this time, I also wanted to gain the perspectives of the HCPs who work to support pwMND, to investigate their experiences, current practices, and attitudes to cognitive and behavioural changes in MND. Consequently, I designed and implemented a survey, the findings of which are described in Chapter 6. The findings from this survey were used to guide the subsequent HCP interviews, as described in Chapter 7.

As shown in Chapter 3, the findings of the scoping review revealed that there is no published literature that has explicitly explored the interaction of dysphagia and cognitive and behavioural changes in MND (Francis et al., 2021). Given this, and prior to conducting the HCPs survey, I wanted to extend the literature search to better understand what approaches are available for managing cognitive and behavioural changes in MND more broadly. Therefore, the next chapter (Chapter 5) details a scoping review that was conducted in 2023, which synthesised the literature regarding management strategies for cognitive and behavioural changes in MND.

CHAPTER 5: EXPLORING CLINICAL MANAGEMENT OF COGNITIVE AND BEHAVIOURAL DEFICITS IN MND. A SCOPING REVIEW

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This paper was co-authored, and the contribution of each author is as follows: [5][6]

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Research design, 70%, Data collection and analysis 80%, Writing and editing 70%

Associate Professor Stacie Attrill
Research design, 10%, Data collection and analysis 10%, Writing and editing 10%

Ratko Radakovic
Research design, 5%, Data collection and analysis 5%, Writing and editing 5%

Professor Sebastian Doeltgen
Research design, 15%, Data collection and analysis 15%, Writing and editing 15%

[5] This chapter was peer reviewed and published online in *Patient Education and Counseling* and subsequently in print.

[6] References within this chapter have been reformatted to meet the requirements of this thesis.

Introduction

Motor Neurone Disease (MND) is an umbrella term used for a class of heterogenous neurodegenerative, sometimes rapidly progressing, terminal diseases, of which the most common phenotype is ALS (Wood-Allum & Shaw, 2010). Although previously thought to be only associated with degeneration of primary motor cortical circuits (Brennan et al., 2022; Lillo & Hodges, 2010), MND is increasingly recognised as a multi-system disease (Brennan et al., 2022; Spalloni & Longone, 2016), that can also include changes to cognition and behaviour (Lillo & Hodges, 2010; Neary et al., 2000). Clinically, cognitive and behavioural deficits are linked to poorer patient outcomes (Govaarts et al., 2016), reduced treatment uptake (N. H. Martin et al., 2014), reduced adherence to interventions (N. H. Martin et al., 2014; Phukan et al., 2007) and shortened life expectancy (Goldstein & Abrahams, 2013; Henstridge et al., 2018; Strong et al., 2017). In addition, these symptoms have been shown to increase caregiver burden (Lillo et al., 2012; Phukan et al., 2007; Watermeyer et al., 2015). These negative impacts are important to consider in management, as cognitive and behavioural changes associated with MND is thought to occur in up to 50% of people living with the disease (pwMND), of which 35% experience mild-moderate symptoms, and 15% will meet the diagnostic criteria for fronto-temporal dementia (Barson et al., 2000; Lillo & Hodges, 2009; Portet et al., 2001).

Literature referring to cognitive and behavioural changes in MND prominently describes symptom presentation and how this should be assessed. For example, clinically, cognitive deficits in MND may manifest as changes to executive functioning including planning, problem solving, decision making, language and social cognition (Abrahams et al., 2005; Goldstein & Abrahams, 2013), whereas behavioural deficits may manifest as apathy, rigidity and disinhibition (Raaphorst et al., 2012). Current clinical guidelines for MND recommend that routine care encompasses a consideration of a pwMND's capacity, screening for cognitive and behavioural changes, attaining informed consent, as well as healthcare

professional training for cognitive changes in MND (Andersen et al., 2012a; Miller et al., 2009; National Institute for Health and Care Excellence, 2016). In some instances, complete neuropsychological assessment is necessary, with further linkage and reference to dementia specific guidance (National Institute for Health and Care Excellence, 2016). However, these guidelines do not extend to specific recommendations or strategies to support pwMND who experience cognitive and behavioural symptoms, instead providing general considerations relating to tailoring care, communication, advanced decision making and planning for end of life.

Early identification of cognitive and behavioural deficits, regular ongoing assessment, and specific strategies for supporting HCPs who provide care for pwMND, and their families are critical to facilitate an understanding of the disease and improve the lived experience of patients and their families. This is particularly important in the context of person-centred care (Australian Commission on Safety and Quality in Health Care, 2023), where care planning is conducted in partnership with pwMND and their family members, and management respects and incorporates their personal needs and wishes. For pwMND, cognitive and behavioural skills influence their capacity to actively participate in their own care planning and decision making for medical interventions, advanced care planning and end of life care conversations. As such, an awareness by HCPs, patients and their families of cognitive and behavioural change may better support pwMND and their carers to cope and lessen care-giver burden. Whilst there are now cognitive and behavioural screening and assessment tools validated for use in MND (Simon & Goldstein, 2019; S. Woolley, M. K. York, et al., 2010), limited evidence exists regarding the specific strategies that may guide HCPs to manage cognitive and behavioural changes once identified. This may lead to HCPs, pwMND and their carers managing cognitive and behavioural changes in an *ad hoc* manner, or these not being proactively addressed and managed at all.

In parallel, there is increasing recognition that approaches for cognitive impairment should include strategies to facilitate and preserve a person's capacity to participate in decision-making, especially in relation to factors concerning care planning and the nature of their disease (Hegde & Ellajosyula, 2016). PwMND concurrently present with progressive communication impairment as a consequence of the loss of the motor neurons for speech, voice and respiration. Cognitive skills underpin the capacity to communicate, understand communication loss and compensate for this loss. Therefore, management of cognition and behaviour for pwMND has critical implications for both supporting people to participate in decision making regarding their disease and being able to functionally communicate their preferences and decisions.

As little empirical evidence exists to guide pwMND, carers and families and HCPs, a scoping review was conducted to identify and collate strategies specific to managing cognitive and behavioural changes in MND, addressing the following research question:

How are cognitive and behavioural changes associated with MND managed?

Materials and methods

We conducted a Scoping Review using the Arksey and O'Malley (2005) framework, as this allowed exploration of peer-reviewed and grey literature, using a rigorous process to identify and map the available evidence (Munn et al., 2018). This approach also allowed the inclusion of clinical guidelines, information produced by MND support services, patient information sheets and similar documents. It was important to capture the grey literature accessible to pwMND, their families and HCPs to allow analysis of the full range of information and resources available to guide clinical practice (Paez, 2017).

A medical librarian assisted with designing the master search. Initial search terms related to MND AND cognition & behaviour AND management (see Table 4) were included. The search was restricted to English only and a publication date from 2010 onwards to capture

contemporary care approaches to intervention for MND, with a research focus of rehabilitation and care strategies rather than disease cure. As our aim was to broadly investigate management strategies specific to the full spectrum of cognitive and behavioural deficits in MND, the term frontotemporal dementia was not included as a specific search term. However, papers that commented on frontotemporal dementia but were additionally specific to MND were included, i.e., frontotemporal dementia was not an exclusion criterion. The master search was translated and run across Embase, Medline, Psychinfo and Emcare in October 2022. Further snowball searching of the reference lists of included studies resulted in one additional study (Miller et al., 2009) identified for inclusion. The search was translated and run in Google Scholar and Google in October 2022 to access the relevant grey literature.

Table 4.

List of search terms related to management of cognitive and behavioural changes in Motor Neurone Disease (MND)

Search terms relevant to 'Motor Neurone Disease'	Search terms relevant to 'Cognitive/behavioural deficits'	Search terms relevant to 'Management'
Motor Neurone Disease	Cognition	Mang*
Motor neurone disease	Behaviour*	Strateg*
MND	Behavioural change	Cope
Amyotrophic lateral sclerosis	Apathy	Maintain*
ALS	frustration intolerance	Control
Progressive Bulbar Palsy	insight	deal with
PBP	rigidity	Recommend*
Primary Lateral Sclerosis	impulsivity	Guideline*
PLS	preservation	Practice pathway
Progressive muscle atrophy	executive functio*	Clinical pathway
PMA	disinhibition	Practice guideline
Global MND	compuls*	Strateg*
Flail arm	think*	Cope
flail leg	focus	Maintain*
bulbar onset	concentrate*	Control
spinal onset	inertia	
Lou Gehrig's disease		
Gehrig's disease		
SLA		
Progressive spinal muscular atrophy		
PMSA		

Note: *Search terms used across Embase, Medline, Psychinfo and Emcare.*

For the interest of clarity peer-reviewed literature are referred to as studies, whereas grey literature findings are referred to as documents.

The initial search of the peer-reviewed literature yielded 1989 studies, which were uploaded into the online software platform Covidence™ for screening. Of these, 299 were identified as duplicates and removed. Title and abstract screening was conducted by two members of the research team, who were blinded to each other's ratings. Of the 1690 studies screened, 1648 did not meet the inclusion criteria and were removed. Consequently, 42 studies underwent blinded full text review by two reviewers, with conflicts resolved by consultation with a third reviewer. Of these, 26 studies met the inclusion criteria (Figure 18).

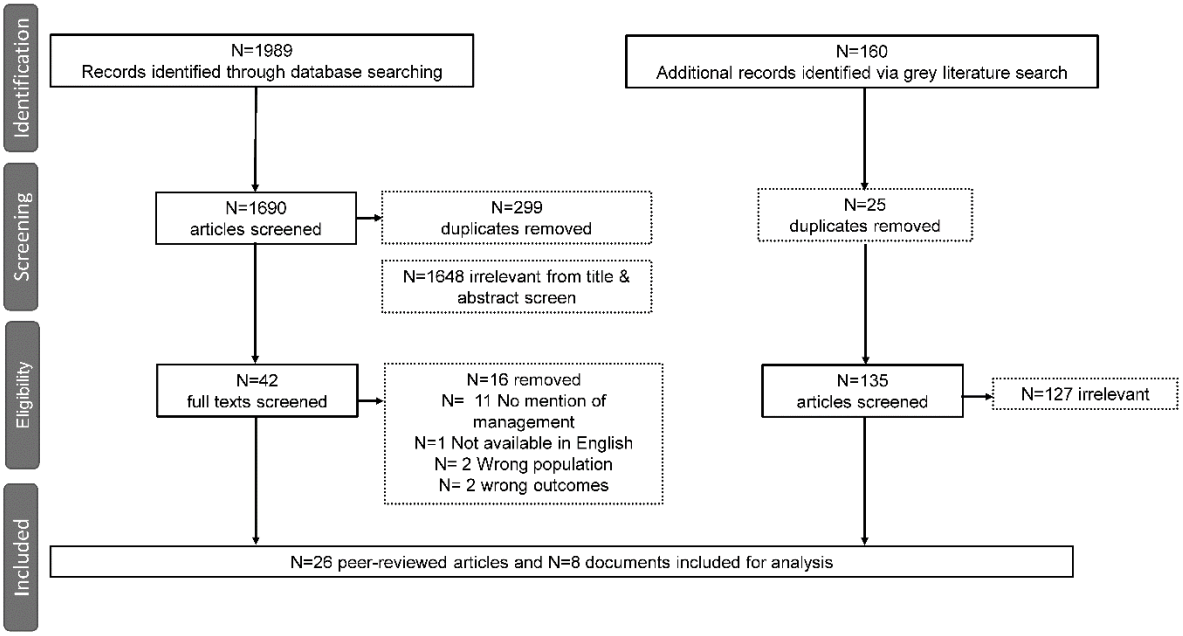


Figure 18: PRISMA literature search
(PRISMA: Preferred Reporting Items for Systematic reviews and Meta-Analyses). **Image created by thesis author.**

A search of the grey literature was conducted across Google Scholar and Google. As redundancy of the search outcomes was reached within the first 160 results, these were uploaded into EndNote and Covidence for analysis and deduplication. Twenty-five peer-reviewed studies were identified as duplicates and removed. Two reviewers who were blinded to each other's ratings screened the remaining 135 search results in Microsoft Excel. Of these, 127 search results did not meet inclusion criteria, with the remaining eight search results included in the analysis. For both peer-reviewed and grey literature, conflicts between raters were resolved by consensus.

A formal grading assessment of individual studies was not conducted, as the aim of this review was to provide an overview of the available evidence rather than comment of the quality of that evidence. A data-charting form was developed in Microsoft Excel by two

reviewers to chart the data of the 26 included peer-reviewed articles and eight included documents. Key characteristics of each study were then extracted using a general inductive approach (Thomas, 2006), which included country of publication, publication year and study type as represented in Figure 19. Additionally, a verbatim copy of text passages that directly related to the research question and search terms was extracted.

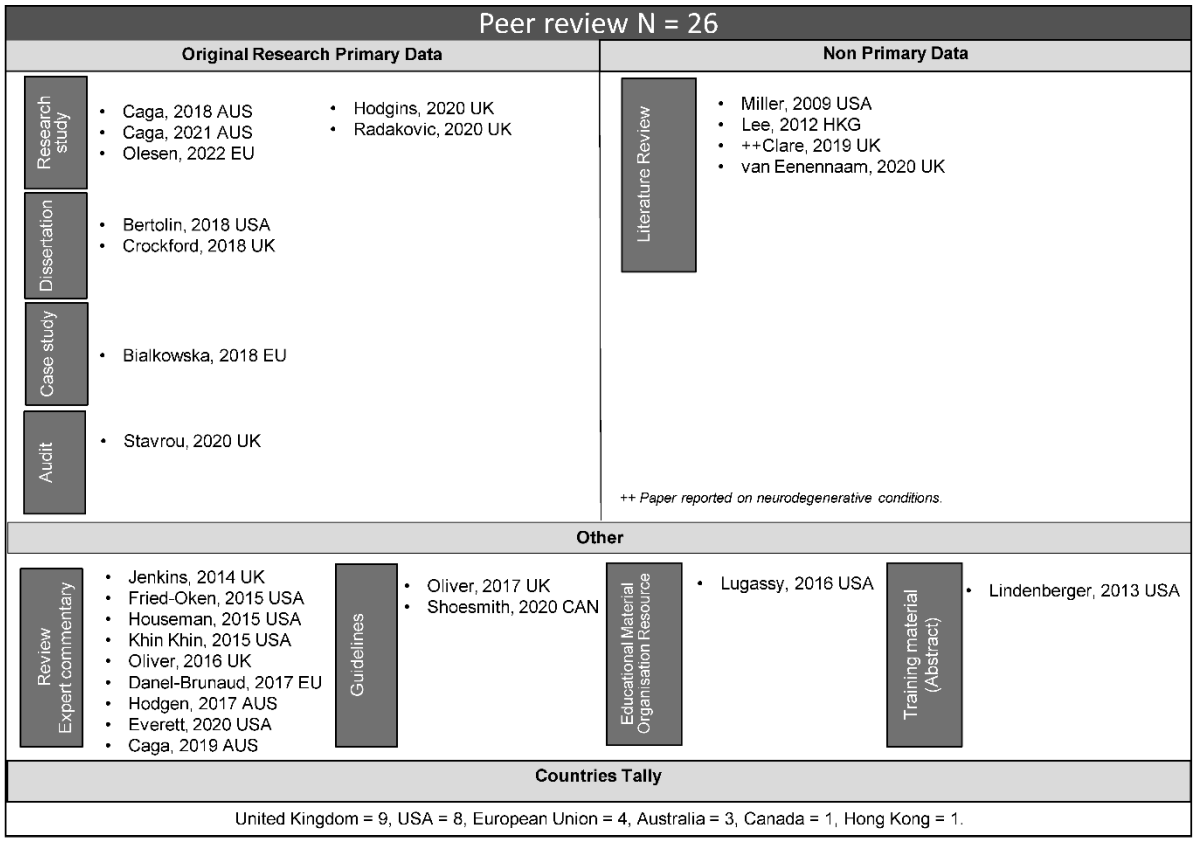


Figure 19: Data charting. Key characteristics of each study were extracted and included country of publication, publication year and study type. Image created by thesis author.

Two members of the research team completed analysis of the extracted qualitative data related to the management of cognitive and behavioural changes. Specifically, the verbatim copy of text passages from both the peer-reviewed literature and documents extracted were discussed, mapped and categorised. Sense checking was conducted by all authors following further conversation and discussion. The themes presented below have been ordered to most closely represent a MND clinical pathway, where review and assessment is

undertaken, education is provided and care planning is implemented (MND Australia, 2017). A summary of the key patterns observed in the grey literature, and how these align with the patterns in the peer-reviewed literature is included in the results section.

Findings

Of the 26 included peer-reviewed studies (Figure 19), nine were original research with primary data. Of these, five were research studies, which included a four-round Delphi model with HCPs working with pwMND (Radakovic et al., 2020), one cross sectional cohort study of 51 caregivers of pwMND (Caga et al., 2018), one cross sectional cohort study of 15 caregivers of pwMND (Olesen et al., 2022), a mixed methods study of 22 MND care services (Hodgins et al., 2020) and a factor analysis study of 39 pwMND with cognitive and behavioural change (Caga et al., 2018). Additionally, there were two dissertations, one case study and one audit. There were four non-primary data studies, which all were forms of literature review. The largest group of studies (N=9) were expert commentary and in addition, there were two guidelines, one abstract, which referenced a training workshop for MND and one organisational resource.

Included peer-review articles were published mostly in the United Kingdom (n=9) and the United States of America (n=8), followed by the European Union (n=3), Australia (n=3), Canada (n=1) and Hong Kong (n=1). All studies are listed by type and referenced in Figure 19.

The eight included grey literature documents (Table 5) were from the United Kingdom (n=5) and the United States of America (n=3). Of these, three were clinical guidelines, from which two aimed to guide healthcare professionals regarding cognitive change and fronto-temporal dementia in MND, and one was a National Institute for Health and Care Excellence (NICE) guideline (National Institute for Health and Care Excellence, 2016), which described assessment and management of MND more broadly. In addition, there were two news articles, a single fact sheet, book and webpage. One of the news articles (Anne Rowling Regenerative Neurology Clinic, 2020) was a summary of an included peer-reviewed article. Whilst this is a duplicate, it was considered important to include to show the breadth of the types of literature available to HCPs and consumers.

Table 5.

Grey literature documents

Article name	Literature type	Published by	Date	Authors	Country	Description of content related to management of cognitive & behavioural change in MND
Cognitive change, frontotemporal dementia and MND	Guidebook for health professionals	Motor Neurone Disease association	July 2021	Abrahams, Goldstein, Snowden, Rohrer	UK	Specific Strategies addressed in Chapter titled Management Strategies. Pg. 27 to Pg. 31
Results published of audit of MND cognitive assessments led by Rowling Scholar	News Article	Anne Rowling Regenerative Neurology Clinic	April 2020	Stavrou	UK	++Provided a summary of the audit research article by Stavrou, 2018 included in peer review search
FYI: ALS, Cognitive Impairment & Dementia	Guide for professionals	ALS Association	Revised in 2014	Rush, Murphy, Lion	USA	Specific strategies addressed titled Recommendations for Routine Management and Care

Article name	Literature type	Published by	Date	Authors	Country	Description of content related to management of cognitive & behavioural change in MND
Supporting people living with motor neurone disease	Factsheet	Royal College of Speech & Language Therapists	Unknown	Unknown	UK	Decision-making: SLTs can contribute to supported decision making and the determination of mental capacity where changes to cognition and behaviour impact on thinking or give the impression of impacting on thinking. This promotes better planning and decision-making and reduces the risk of people being perceived as lacking capacity due to their communication needs not being appropriately supported. Pg 3
Confirmed: ALS Attacks Cognition and Behavior	News page	ALZFORUM	2016	Murphy, Mitsumoto	USA	…caregivers of people with both ALS and behavioural symptoms may require extra support. … HCPs need to know how clearly their patients are thinking so they do not expect them to understand concepts or procedures or make decisions that they are not equipped to make.

Article name	Literature type	Published by	Date	Authors	Country	Description of content related to management of cognitive & behavioural change in MND
Motor neurone disease: assessment and management	Guideline	National Institute for Health and Care Excellence	Updated 2019	Unknown	UK	1.3.1 Be aware that people with MND and frontotemporal dementia may lack mental capacity. Care should be provided in line with the Mental Capacity Act 2005. At diagnosis, and if there is concern about cognition and behaviour, explore any cognitive or behavioural changes with the person and their family members and/or carers as appropriate. If needed, refer the person for a formal assessment in line with the NICE guideline on dementia. Tailor all discussions to the person's needs, taking into account their communication ability, cognitive status and mental capacity. Pg. 9
Living with Motor Neurone Disease: A complete guide	Book	Project Muse	2021	Pender, Pinto-Grau	Ireland	Chapter 4. Will Motor Neurone Disease Affect My Mind? Cognitive and behavioural changes in MND
Dementia in Amyotrophic Lateral Sclerosis	Webpage	WebMD	2021	Melinosky	USA	Treatment of dementia in ALS and other motor neuron diseases focuses on relieving symptoms.

Thematic analysis of the included literature revealed six distinct themes that described a specific feature of cognitive and behavioural changes in MND to manage (Tables 6 to 11). The information contributing to each theme was grouped into either *specific strategies,* if there was an explicit description of the management strategy, or *general recommendations,* if reference was made to an area suggested for management without a specific or detailed management description.

Note, while distinct themes were identified in the analysis, in clinical practice some of these themes are likely to overlap. For example, provision of both education and clear and open communication are likely critical to facilitate carer support.

Theme 1. Cognitive and behaviour assessment

Screening for cognitive and behavioural deficits in MND was discussed in 12 of the 26 included studies (Table 6), suggesting that early identification via screening and regular monitoring is an important part of overall MND management.

The grey literature similarly positioned screening for cognition and behavioural changes in MND as part of managing these symptoms. For example, the publicly available document Cognitive Change, frontotemporal dementia and MND published by the MND Association for the MND Community of Practice is a guide for professionals working in MND and includes practical tips for management. This guide also suggests that timely assessments are important and further adds that this supports understanding of the disease and coping with future changes. The guide incorporates recommendations from the NICE guidelines (National Institute for Health and Care Excellence, 2016) and outlines clear processes for conducting an assessment, with recommendations for neuropsychology involvement if practical. However, in contrast to the peer-reviewed papers, this guide also suggested that specific strategies to manage cognitive and behavioural deficits are required once these symptoms have been identified. In particular, assessment was suggested to inform specific

strategies to support a pwMND in decision making, learning, problem solving, communication and participation.

The NICE (National Institute for Health and Care Excellence, 2016) guidelines suggest that further work is needed to determine if assessing for cognitive and behavioural change in MND improves patient outcomes: *A randomised controlled trial is needed to assess whether formal assessment at diagnosis and/or repeated assessment improves clinical practice, subsequent care of the person and quality of life for the person, their family and carers. (*Pg. 39).

Specific reference is made within both the peer-reviewed and grey literature to the screening tools Edinburgh Cognitive Assessment Screener (Niven et al., 2015) and the ALS Cognitive Behavioural Screen (ALS-CBS) (Woolley et al., 2010) which are both validated for use with this population. Four of the included peer-reviewed studies (Jenkins et al., 2014; Miller et al., 2009) suggested further neuropsychological evaluation may be necessary, but acknowledge limited time and resources make this at times impractical.

In summary, there was consensus amongst the peer-reviewed and grey literature that assessing for cognitive and behavioural changes is an important component of managing this symptom. While the 12 studies included in this theme referred to screening and assessment procedures as important, they were largely silent about management processes beyond assessment.

Table 6.

Theme 1. Cognitive and behaviour assessment

Citation	Strategy	Management description	Enacted by	Recipient
Khin Khin, 2015	SS	A range of comprehensive neuropsychological tests can provide nuanced and specific data on cognitive ability. However, the time and resource-intensive nature of neuropsychological batteries hinders their practical and widespread use in ALS clinics. A more plausible approach would be to detect possible cognitive and behavioural impairment at an early stage via brief screening tests and then to refer patients with positive results for more extensive testing. One practical framework proposed by Strong, and colleagues involved a hierarchical approach outlining the following testing paradigms: a brief screening of 2 to 5 minutes, a more extensive assessment of 5 to 20 minutes, and formal neuropsychiatric testing. As they noted, screening tests cannot be used in formal diagnosis of cognitive ALS-FTD, ALSbi, or ALSci. Pg. 212 By conducting brief cognitive and behavioural screening of patients at regular intervals, those who start to show deficits in these areas can be identified and monitored closely. Pg. 216	HCP	pwMND
Stavrou, 2020	SS	Additional pathways should be developed for cognitive/behavioural screening for pwMND such as: (1) Masterclasses /training days to enhance health care professionals' knowledge in ECAS; (2) Establishment of dedicated ECAS clinics or incorporating psychologists into clinic; (3) Ongoing support and access to neuropsychology services. Neuropsychological intervention helps the MDT manage the particularly complex cases. Pg. 462	HCP	HCP

Citation	Strategy	Management description	Enacted by	Recipient
van Eenennaam, 2020	SS	If due to cognitive impairment/FTD the patient is suspected of lacking decisional capacity to decide whether they want to discuss their prognosis, a cognitive screener like the Edinburgh Cognitive and Behavioural ALS Screen can be used to gain insight into affected cognitive domains. Table 1. Pg 5	HCP	pwMND
Shoesmith, 2020	GR	Screening for cognitive and behavioural impairment should be performed in patients with ALS early in their disease. Table 1. If there is concern about cognition or behaviour at any point, specific assessments should take place with the person and their family members or caregiver, as appropriate (EC). Table 1 (part 5 of 6). Pg. 1458	HCP	pwMND
Crockford, 2018	GR	Recently updated ALS consensus guidelines suggests that all people with ALS be assessed using the ECAS or ALS-CBS. Pg. 61 Cognitive and behavioural screening in MND is important in the management and care of patients and their families, as highlighted by recently updated guidelines from the National Institute for Health and Care Excellence (NICE) Pg.206	HCP	pwMND
Bertolin, 2018	GR	However, the selection and implementation of therapeutic interventions may be significantly impacted by a patient's neuropsychological status, so identification of co-occurring cognitive and behavioural impairment is an essential component of clinical care for this population Pg.54	HCP	pwMND
Oliver, 2017	GR	At diagnosis or if there is a concern about cognition or behaviour explore these areas with the person and their family. If necessary, undertake a formal assessment. Assess for capacity and adjust care accordingly Pg. 320	HCP	pwMND

Citation	Strategy	Management description	Enacted by	Recipient
Lugassy, 2016	GR	The risk of developing cognitive impairment and dementia in ALS necessitates ongoing assessment of capacity for medical decision making in the context of the ongoing goals of care discussions which ALS typically requires Pg. 3	HCP	pwMND
Fried-Oken, 2015	GR	Because ALS affects motor function and often cognitive function as well as speech, patients should be regularly screened for changes that might affect communication. Pg. 75 Early identification of cognitive changes… Screening tools such as the ALS Cognitive Behaviour Screen can monitor cognitive functioning...Pg. 75	HCP	pwMND
Jenkins, 2014	GR	Rapid identification is facilitated by screening tools such as Edinburgh Cognitive Assessment Scoring. Treatment is difficult; neuropsychological testing may clarify domains affected and enable occupational therapists to provide practical strategies. Pg 528	HCP	pwMND
Lee, 2012	GR	Screening of MND patients for co-morbid conditions at diagnosis and at regular intervals is recommended by the American Academy of Neurology Practice Parameter. Pg. 53-54	HCP	pwMND
Miller, 2009	GR	Screening tests of executive function may be considered to detect cognitive impairment in patients with ALS prior to confirmation with formal neuropsychological evaluation. Pg. 1231	HCP	pwMND

Note: SS = Specific Strategy. Bold rows denote a specific strategy related to managing cognitive and behavioural change in MND. GR = General recommendation. Non bolded rows denote a general recommendation related to managing cognitive and behavioural change in MND.

Theme 2. Providing education

Data from eight studies comprised the theme, ‘Providing education’, related to managing cognitive and behavioural change in MND, of which three identified *specific strategies.* Two studies (Danel-Brunaud et al., 2017; Houseman & Kelley, 2015) described providing education to the caregiver about the neurological changes that can occur in MND, which manifest as changes to behaviour to avoid misunderstandings between the pwMND and their caregiver. The third study (Radakovic et al., 2020) suggested that education should include common presentations of cognitive and behavioural change in MND to support HCPs and caregivers to recognise these. The remaining studies in this theme made *general recommendations* by suggesting that it is necessary to provide education, but without providing details that could guide HCPs (Table 7). Of note, two studies referred to literature about dementia-related literature (Caga et al., 2021; Radakovic et al., 2020) generating parallels in providing caregiver training for cognitive and behavioural change in MND.

The theme of Providing education was also prominent in the included grey literature and added specific suggestions that complement the *general recommendations* that were prominent in the peer-reviewed literature. For example, within Cognitive change, frontotemporal dementia and MND (Motor Neurone Disease Association, 2022) (Pg. 9), HCP education included a description of functional deficits related to cognitive and behavioural change in MND and identified specific strategies to manage these (Pg.24).

In summary, the provision of education was described as an important component in the management of cognitive and behavioural change in MND. However, the peer-reviewed literature mostly suggested that this education be targeted at the carer as the recipient, with only one study explicitly suggesting that education be provided to the pwMND (Caga et al., 2021).

Table 7.

Theme 2. Providing Education

Citation	Strategy	Management Description	Enacted by	Recipient
Radakovic, 2020	SS	…educational material on common cognitive and behavioural impairments (and how they might be observed) would be beneficial for inclusion in the toolkit for both the healthcare professional and the family members or caregivers. Pg.20 Information/education for caregivers/families and staff about impairment and impact Pg. 21	HCP	HCP & Caregiver
Danel-Brunaud, 2017	SS	Education and counselling on the neurological bases of behavioural symptoms can help caregivers (and especially family members) to avoid misunderstandings such as ‘‘he’s (she’s) doing it on purpose’’ and affective frustration (‘‘he (she) is ungrateful and does not love me anymore Pg. 305	HCP	Caregiver
Houseman, 2015	SS	Educate the caregiver about CI/FTD, in particular, explain the organic nature of the illness (that it is an illness, person cannot help his or her CI/behavior Pg.125 Address safety and environmental concerns, including medication management, driving, managing finances, use of power tools/equipment, keeping anything harmful out of sight/reach. Pg. 126 Address (possible) behavioural concerns by using a calm voice, giving one-step directions, avoid trying to reason with the patient, supervising activities such as eating and bathing, finding and utilising soothing activities. Pg. 126	HCP	Caregiver

Citation	Strategy	Management Description	Enacted by	Recipient
Caga, 2021	GR (Derived from literature about populations with dementia).	patient and caregiver education session initiated by the multidisciplinary team shortly after diagnosis of cognitive and behavioural involvement may be timely. Education tailored to specific cognitive, and emotion related ALS perceptions may focus on managing extremely distressing perceptions about the impact of the ALS on functional capacity and quality of life. This may also be beneficial prior to the implementation of supportive interventions to minimise functional disability and optimise intervention uptake and adherence Pg. 5 …there have been no interventions for cognitive or behavioural symptoms systematically tried to date. General strategies to manage cognitive and behavioural symptoms based on the dementia literature have been proposed. First and foremost is patient (depending on their level of insight) and caregiver education provided by a multidisciplinary team. Pg. 5	HCP	pwMND & Caregiver
Radakovic, 2020	GR (Derived from literature about populations with dementia).	Other approaches were more personalised in terms of providing more general support, education and information for family members or caregivers about cognitive and behavioural impairment, which are also often observed in management of dementia, for example. Pg. 21-22	HCP	Caregiver
Hogden, 2017	GR	Expert consensus is that care provision in ALS should include interventions directed toward the management of cognitive and behavioural impairment in ALS, including educational and support services for family carers on how best to support and care for ALS patients who have cognitive or behavioural impairment. Pg. 211	HCP	Caregiver
Van Eenernnaam, 2020	GR	Discussing estimated survival with the patient's family, if they want to know, can still be important as they will have to take into account a poorer prognosis due to cognitive impairment Pg. 7	HCP	Caregiver

Citation	Strategy	Management Description	Enacted by	Recipient
Caga, 2018	GR (Derived from literature about populations with dementia).	In dementia, education focusing on training caregivers to modify their interactions with patients appears to be highly beneficial for managing behavioural symptoms Pg. 603	HCP	Caregiver

Note: SS = Specific Strategy. Bold rows denote a specific strategy related to managing cognitive and behavioural change in MND. GR = General recommendation. Non bolded rows denote a general recommendation related to managing cognitive and behavioural change in MND

Theme 3. Advanced care planning

Five studies (Table 8) spoke to the theme of advanced care planning (ACP) specifically related to the management of cognitive and behavioural deficits in MND. Of these, two studies described *specific strategies.* One suggested early ACP discussions be presented to pwMND and their families as part of routine practice, as cognitive impairment is common in MND. The authors noted that this approach may address concerns related to the sensitivity of the topic, and further suggested that families welcome these discussions (Everett et al., 2020). The other study gave *specific strategies* regarding the appointment of a legal medical power of attorney to give instructions in the event of impaired communication.

The remaining four studies (Table 8) made *general recommendations* related to ACP and identified that discussions would be best placed to occur earlier in the disease due to the complex interplay between cognitive and behavioural change in MND, and the pwMND's capacity to participate in decision-making as the disease progresses. The included studies did not explicitly relate ACP as supporting the complex relationship between cognitive and behavioural changes and communication function in pwMND. One study (Oliver, 2016) noted that if cognitive decline is identified, decision making related to care may not directly involve the pwMND.

ACP was also discussed and recommended in the grey literature in particular reference to cognitive changes for pwMND. For example, an Advance Decision to Refuse Treatment (ADRT) approach (Motor Neurone Disease Association, 2022) is described (Pg. 29), which allows a pwMND to be pro-active in their own care planning, should capacity or communication become impaired with disease progression.

In summary, ACP is described as an important part of management related to cognitive and behavioural change in MND, with most resources referencing the need for this to be conducted early in disease progression.

Table 8.

Theme 3 Advanced care planning

Citation	Strategy	Management description	Enacted by	Recipient
Everett, 2020	SS	ACP in ALS should start early…and should continue throughout the disease course as treatment preferences often change with symptom progression. Many HCPs worry that patients and families may feel that discussions about ACP happen too early, although framing ACP as a 'normal part of practice that is done with all patients'' can eliminate that concern. Pg. 843	Not provided	Not provided
Houseman, 2015	SS	Advise of the need for the person with ALS to complete his or her Advanced Directive and Living Will. The patient should also identify and ensure a legal medical power of attorney; this person would speak on the behalf of the person with ALS (re: future medical care, if the person with ALS is unable to do so). Pg. 126	HCP	pwMND
Shoesmith, 2020	GR	Because the presence of frontotemporal dementia negatively affects survival, ACP should be done early in the disease (EC). Table 1. Pg. E1458	Not provided	Not provided
Crockford, 2018	GR	…decisions about medical care should be made early in the disease to maximise the potential for capacity Pg. 248	Not provided	Not provided
Oliver, 2016	GR	If patients are to be included in decision making, it may be necessary to consider advance care planning, such as an advance directive or the definition of a proxy for decision making, to ensure that their wishes are known and can be respected Pg. 67	HCP	pwMND

Citation	Strategy	Management description	Enacted by	Recipient
Khin, 2015	GR	…advance directives and surrogate decision making can be used to preserve patients' choices so that they can give directions for future medical care and have their wishes honoured in the event of incapacity. Pg. 215	Not provided	Not provided

Note: SS = Specific Strategy. Bold rows denote a specific strategy related to managing cognitive and behavioural change in MND. GR = General recommendation. Non bolded rows denote a general recommendation related to managing cognitive and behavioural change in MND.

Theme 4. Adaptation of care

This theme relates to adaptations of routine care, which are recommended when cognitive and behavioural change has been identified. Of the eight studies included in this theme, four provided *specific strategies* to guide HCPs, including specific strategies to manage deficits: using a supportive communication style, addressing safety concerns, supporting decision making and adapting the environment (Table 9). The remaining studies provided *general recommendations* that the care plan should be adapted or adjusted but without describing specific information (Table 9).

The grey literature provided more *specific strategies* to guidance related to adapting care than the peer-reviewed literature. Specifically, the MND Association guide (pages 27 to 35) (Motor Neurone Disease Association, 2022) outlines strategies for HCPs or caregivers to facilitate them to simplify decision making, help a pwMND to problem solve, support activities of daily living and recognise and manage inappropriate behaviours. Of note, this document referenced literature that describes the patterns and presentations of cognitive and behavioural change in MND, as well as various screening tools for assessment and the NICE (National Institute for Health and Care Excellence, 2016) guidelines. However, the literature that was drawn from did not include research that investigated management strategies of cognitive and behavioural change in MND.

Further, the NICE (National Institute for Health and Care Excellence, 2016) guidelines included specific recommendations for managing other symptoms of MND. For example, details are provided on managing nutrition, gastrostomy or respiratory function; however, management of the symptom of cognitive and behavioural change is not specifically described. Instead, the reader is referred to links to the Mental Capacity Act and the NICE guidelines on dementia (Managing symptoms Section 1.8 Pg. 16-33).

In summary, the need to adapt the approach to care of pwMND who present with cognitive and behavioural change is clearly recognised in both the peer-reviewed and grey literature.

While several resources provided specific guidance to pwMND, carers and HCPs, it is also noted that these resources draw from evidence generated from outside of the MND context.

Table 9.

Theme 4. Adaptations of care

Citation	Strategy	Management description	Enacted by	Recipient
Radakovic, 2020	SS (Derived from literature about populations with dementia).	Structured support approaches (Taken from Table.2) Pg. 21 Strategies for managing the deficits or issues identified: Adapting the environment (e.g., more/increasing structure, less distraction, safety measures) Breakdown tasks into small manageable steps Verbal interaction Distraction Routine Physical focus Behavioural approach Person centered. Family work/systemic approach Acceptance and commitment therapy Personal support approaches Compassionate communication Increased time Make suggestions and let person come to own conclusions …evidence-based techniques, such as cognitive stimulation therapy and behavioural management techniques, as well as explicit mention of acceptance and commitment therapy. These therapies have been found to be effective in dementia… Pg. 21 …the application of these approaches occurs at variable points in the MND care pathway, due to lack of standardised practice or guidance in relation to interventions. Pg. 22	HCP	HCP Carer pwMND
Houseman, 2015	SS	Address safety and environmental concerns, including medication management, driving, managing finances, use of power tools/equipment, keeping anything harmful out of sight/reach Pg. 126	HCP Caregiver	pwMND

Citation	Strategy	Management description	Enacted by	Recipient
Oliver, 2016	SS	…simple decision between two clear options to facilitate the patient's involvement Pg. 67	HCP Caregiver	pwMND
Clare, 2018	SS (Derived from literature about populations with neurodegenerative conditions).	Individualised, goal-oriented interventions for behaviour are based on an individual formulation and on an understanding of the antecedents, consequences and functions of the given behaviour, and can help to reduce the impact of behavioural changes and hence support everyday functioning, relationships and social interactions. Techniques include introducing distraction and controlling triggers, as well as implementing environmental modifications, aids and adaptations. Pg. 13	HCP	Not detailed
Stavrou, 2020	GR	care planning should be adapted for people with cognitive impairment. Pg 460	HCP	pwMND
Crockford, 2018	GR	Further outreach and resources may be required in order to maximise the inclusion of cognition and behaviour in the clinical care management of patients with ALS Pg. 248	Not detailed	Not detailed
Oliver, 2016	GR	Assess for capacity and adjust care accordingly. Tailor discussions to the person's needs, taking into account their communication ability, cognitive status and mental capacity Pg. 320	HCP	pwMND
Miller, 2009	GR	There are insufficient data to support or refute the impact of cognitive and behavioural impairment on management in ALS Pg. 1231	Not detailed	Not detailed

Note: SS = Specific Strategy. Bold rows denote a specific strategy related to managing cognitive and behavioural change in MND. GR = General recommendation. Non bolded rows denote a general recommendation related to managing cognitive and behavioural change in MND.

Theme 5. Communication

The theme Communication relates to strategies available to all stakeholders to facilitate communication with a pwMND who also presents with cognitive and behavioural changes. Of the six studies included in this theme (Table 10), one provided *specific strategies* to support HCPs to communicate with pwMND by delivering information compassionately and with sufficient time to allow the pwMND to process the information (Radakovic et al., 2020).

In the remaining five studies (Table 10), the *general recommendations* made suggestions that communication can be difficult with a pwMND who has cognitive and behavioural changes and communication should be adjusted. However, specific strategies to guide HCPs were not discussed. One way of adjusting communication was described as using alternative and augmented communication (AAC) to support impaired communication (Table 10); However, cognitive decline was also suggested to impact the use and uptake of such communication tools (Fried-Oken et al., 2015; Shoesmith & Strong, 2006)

Table 10.

Theme 5. Communication

Citation	Strategy	Management description	Enacted by	Provided to
Radakovic, 2020	SS	(Taken from Table.2) Pg. 21 - Compassionate communication - Increased time - Make suggestions and let person come to own conclusions …breaking down tasks into manageable small steps, verbal interaction and routine were noted as the best approaches. Pg. 19 Simplify decision-making (e.g., limit choices and reduce open-ended questions) Support problem-solving (e.g., specify the topic being discussed, prompt if needed) Clarify complex information (e.g., breakdown information, explain terminology that is unfamiliar) Pg. 19	HCP	pwMND
Shoesmith, 2020	GR	The choice of communication devices should be tailored to the patient's needs and abilities. Patients with cognitive impairment may need individualised strategies for communication. …the challenges of intervention compliance with cognitive or behavioural impairment should be discussed with the patient and family before deciding to proceed with an intervention. Table 1 Pg. E1458	HCP	pwMND
Hodgins, 2020	GR	…facilitating communication between family members to promote understanding and manage stressful situations… Pg. 97	HCP	pwMND & Caregiver
Everett, 2020	GR	…behavioural changes make communication difficult even if patients maintain decision-making capacity. Pg. 843		
Oliver, 2017	GR	Tailor discussions to the person's needs, taking into account their communication ability, cognitive status and mental capacity Pg.320	HCP	pwMND

Citation	Strategy	Management description	Enacted by	Provided to
Fried-Oken, 2015	GR	…early instruction in multiple forms of AAC, including simple, low-tech strategies, can help avoid such difficulties. Pg. 75 Some individuals with cognitive impairment may reject AAC intervention. Pg. 75	HCP	pwMND & Caregiver

Note: SS = Specific Strategy. Bold rows denote a specific strategy related to managing cognitive and behavioural change in MND. GR = General recommendation. Non bolded rows denote a general recommendation related to managing cognitive and behavioural change in MND.

Theme 6. Carer Support

The theme Carer Support outlines strategies aimed at supporting a carer of a pwMND to manage cognitive and behavioural change. There were eight studies included in this theme (Table 11), of which two described *specific strategies* addressing support for carers to manage these changes. An intervention program called Embrace (Olesen et al., 2022) described cognitive and behaviour support aimed at improving the experience for carers of pwMND. The second study (Radakovic et al., 2020) detailed the provision of carer support to include education to understand this symptom of the disease and enabling carers to express themselves and take respite. These *specific strategies* oriented carers to recognise that the nature of cognitive and behavioural changes in MND may increase their need to be proactive in their own self-care, respite, and support needs.

The remaining five studies made the *general recommendation* that carer support should be part of managing cognitive and behavioural change in MND (Table 11), however, without providing specific strategies to guide HCPs to enact this.

The theme of Carer Support also appeared in the grey literature, with the MND Association Guide (Motor Neurone Disease Association, 2022) suggesting further *specific strategies* that allow carers to understand and cope with cognitive and behavioural changes. Additionally, the guide suggested that the needs of the carer should be a part of the overall assessment process (Supporting carers and family members, Pg. 26) in an effort to alleviate care-giver burden.

The NICE (National Institute for Health and Care Excellence, 2016) guideline highlighted an additional perspective that was absent from the peer-reviewed literature, suggesting that permission should first be sought from the pwMND prior to consultation with family members or caregivers.

In summary, the importance of supporting the carer of a pwMND with cognitive and behaviour change was recognised in both the peer-reviewed and grey literature. A small number of resources are available to guide HCPs in supporting and advocating for carers, including the provision of carer education, assessing carer needs and empowering carers to advocate for their own care and support needs.

Table 11.

Theme 6. Carer support

Citation	Strategy	Management description	Enacted by	Recipient
Olesen, 2022	SS	EMBRACE intervention, a 4-month online program aimed at supporting the ability of caregivers of PALS/Cis to handle everyday challenges related to the care of PALS/Cis Pg. 2 …new online palliative rehabilitation program (EMBRACE), a blended learning program developed for caregivers of PALS/Cis. Pg. 2	HCP	Caregiver
Radakovic, 2020	SS	Support the family/caregiver (e.g., support the family/caregiver in understanding cognitive and behavioural symptoms, expressing themselves to the patient, taking respite). Pg. 19	HCP	Caregiver
Caga, 2019	GR	Comprehensive management of cognitive and behavioural symptoms not only promotes holistic care of patients but would also further enhance caregiver's psychological well-being… Pg. 4	HCP	pwMND
Hodgins, 2020	GR	Interventions mentioned by psychologists included behavioural interventions-working with carers to implement strategies for managing challenging behavior; relational/family interventions-facilitating communication between family members to promote understanding and manage stressful situations… Pg. 97	HCP	Caregiver

Citation	Strategy	Management description	Enacted by	Recipient
Crockford, 2018	GR	Clinically, it may be necessary to consider intervention programmes for caregivers to alleviate the impact of neuropsychological impairment, particularly early in the disease course. Pg. 131	HCP	Caregiver
Hogden, 2017	GR	Expert consensus is that care provision in ALS should include interventions directed toward the management of cognitive and behavioural impairment in ALS, including…. support services for family carers on how best to support and care for ALS patients who have cognitive or behavioural impairment. In this context neuropsychologists make an important contribution to the care of ALS patients and their caregivers Pg. 211	HCP	Caregiver
Danel-Brunaud, 2017	GR	Education and counselling on the neurological bases of behavioural symptoms can help caregivers (and especially family members) to avoid misunderstandings such as “he’s (she’s) doing it on purpose” and affective frustration (“he (she) is ungrateful and does not love me anymore. Pg. 305	HCP	Caregiver
Caga, 2018	GR (Derived from literature about populations with dementia).	…. strategies for ALS caregivers to manage apathy include cognitive behavioural strategies to ensure realistic expectations about a patient’s capacity to engage in activities as well as participation in alternative activities Pg. 60	HCP	Caregiver

Note: SS = Specific Strategy. Bold rows denote a specific strategy related to managing cognitive and behavioural change in MND. GR = General recommendation. Non bolded rows denote a general recommendation related to managing cognitive and behavioural change in MND.

Discussion and conclusion

Whilst it is now recognised that cognitive and behavioural symptoms often form part of the collection of MND characteristics, less is known about how these symptoms should be managed and how stakeholders are supported in this process. This scoping review synthesised the information and resources available to stakeholders and identified six themes that are organised as a taxonomy of management strategies to guide HCPs, carers, pwMND and researchers. These themes include i. Assessment, ii. Education, iii. Advance Care Planning, iv. Adaptation of Care Plan, v. Communication and vi. Carer Support.

The recommendations drawn from the themes related to:

- the use of the ECAS or ALS CBS for early screening and as part of regular and ongoing management.
- simplifying communication with people identified to have cognitive and behaviour change to support understanding.
- early instigation of advanced care planning to allow for changes in capacity.
- providing education which focusses on the neurological basis behind the functional changes.
- carer support to alleviate misunderstandings and reduce caregiver burden.

In general, it was noted that the majority of studies mentioned *general recommendations* relating to each theme, with far fewer studies providing specific strategies or pathways for managing this symptom of MND.

This review identified that most literature addressing management of cognitive and behavioural changes in MND is derived from descriptive reviews and expert opinion. For example, nine of the 26 studies were classified as opinion papers with only five employing rigorous exploratory methods. This was also reflected in the grey literature, with documents written by experts in the field with limited reference to any primary data for this population.

Some literature that describes managing cognitive changes in dementia was applied to MND (Sadowsky & Galvin, 2012). This association likely resulted from a lack of primary data that explicitly investigates or informs development of strategies for managing cognitive and behavioural deficits specific to MND. Further, much of the primary data about MND is generated through research that has excluded pwMND who present with cognitive and behavioural changes. As up to fifty percent of pwMND experience some change to cognition (47), existing research is potentially lacking validity for a large proportion of the overall MND population (Francis et al., 2021). As these symptoms are known to result in poorer intervention outcomes and prognosis for pwMND (Naomi H Martin et al., 2014; Nguyen et al., 2021), there is an urgent need for high-quality research to a) identify the most appropriate strategies to manage cognitive and behavioural deficits in MND and b) evaluate the benefits of these strategies for pwMND, carers, HCPs and the health care system more broadly.

It is acknowledged that given the debilitating and often rapidly progressing nature of MND and the severe impact this disease has on functioning and independence, it is likely that the physical, or seen, symptoms of MND may take precedence in clinical management and in research. However, as part of this approach, there are decisions and personal preferences for treatments that need to be considered within a person-centred, choice-based and holistic approach to care. There is evidence to suggest that the presence of cognitive and behavioural changes impacts on treatment uptake (7), for example refusal of non-invasive ventilation to manage sleep-disordered breathing (Dorst & Ludolph, 2019) or gastrostomy to manage nutritional and fluid intake (Mazzini et al., 1995). To date, these intervention strategies have not been discussed in the context of cognitive decline**.** Developing an understanding of the cognitive abilities of the pwMND and having available evidence about a range of management approaches, enables HCPs to partner more effectively with pwMND and their families to jointly identify and then apply evidence-based strategies that are most suited to their needs and individual context. Broadening the care team to include not only

HCPs with intervention-specific knowledge, but also experts with an awareness of how cognitive and behavioural change may impact on intervention implementation. This may facilitate pwMND and their families to have greater understanding of the changes they are experiencing, how these changes may be impacting on their decisions, and give them agency in managing these pro-actively.

In this context, early identification of cognitive and behavioural changes is likely a critical factor, and this was a predominant theme of the included literature. This is particularly important in light of emerging evidence suggesting that cognitive and behavioural deficits may, in some cases, precede motor function change in MND (Mioshi et al., 2014; National Institute for Health and Care Excellence, 2016) The provision of assessment may be limited due to underfunding, inadequate pathways for care when impairments are detected, and importantly, the lack of specific guidelines or techniques to guide practice when impairments are detected or observed. To ameliorate this, early identification of cognitive and behavioural changes may permit pwMND to express and discuss their wishes prior to motor (and further cognitive) decline that limits communication.

It is unsurprising that communication was identified as a key theme related to cognitive and behavioural management in MND, given the debilitating and direct impact of the disease on communication function. With disease progression, pwMND may lose the ability to speak intelligibly due to loss of laryngeal, oropharyngeal and diaphragmatic motor control. However, it is also important to consider that cognitive function underpins effective communication skills, and it has been shown that language impairments such as deficits in linguistic processing also occur in pwMND (Sbrollini et al., 2022; Taylor et al., 2013). In addition, cognitive function underpins capacity to learn techniques that may be implemented to support communication loss, such as alternative communication strategies **(AAC)** or devices. Therefore, even in the early stages of the disease, pwMND may need support to understand disease management choices and subsequently communicate their preferences

for ongoing care. Current approaches to AAC would benefit from further consideration of how to optimise communication supports for people with cognitive and behavioural change in MND. Importantly, the overlap of motor and cognitive function enabling effective communication underscores the need to determine early if communication of individual preferences maybe be impaired by a loss of speech motor function or if cognitive impairment is impacting on a pwMND's ability to understand and weigh up options and access the language to express choices.

As a basic human right, the Convention on the Rights of Persons with Disabilities (CRPD) (United Nations, 2006) establishes that people living with a disability are capable of making decisions which best suit their individual beliefs and circumstances and that it is the role of support providers to gain informed consent and adapt care for the person with disability to be able to exercise this right. In addition, person centred care models are based on the understanding of the wishes and preferences of the individual. Seemingly in contrast to this, many of the included general recommendations in this review suggest that conversations take place between the HCPs and the carer of a pwMND regarding management of cognitive and behavioural changes that may be present. This suggests that the carer is responsible for managing the cognitive and behavioural changes independently of the pwMND, and highlights that the complex care decisions of pwMND who present with cognitive and behavioural changes may occur without their involvement. Similarly, the literature included in the theme of Advanced Care Planning reflects an approach of doing to or around the pwMND instead of their active inclusion in educational discussions about cognitive and behavioural change or decision-making processes. Across all studies included in this review, only one source (National Institute for Health and Care Excellence, 2016) suggested it was appropriate to first gain consent from a pwMND to permit health care professionals to undertake discussions about their care with families or caregivers. This underpins the need for research that informs strategies that enable pwMND who experience

cognitive and behavioural symptoms to participate meaningfully and effectively in all decision-making regarding both care and broader life choices.

As such, this review is a call to arms for research and clinical practice to further evaluate and consider the impact of cognitive and behavioural changes in the person-centred support of the pwMND and their carers. In particular, research exploring shared decision-making frameworks for use in practice with pwMND who experience cognitive and behavioural symptoms may facilitate partnership with health HCPs that ultimately improves the understanding and experiences of affected individuals and their families. HCPs who are skilled in facilitating this process, and who understand and can assist with managing cognitive and behavioural changes in MND, have the opportunity to enable true partnerships with patients and their carers. Similarly, recent research endeavours have seen technological advancements, such as brain computer infaces, that make it possible for pwMND to communicate their preferences until much later in the disease course. Further work is also needed to investigate current clinical practice and to better understand the attitudes of both HCPs and families with MND towards cognitive and behavioural supports, including any barriers that may exist to accessing these (Crockford et al., 2017). As highlighted in the NICE guidelines (National Institute for Health and Care Excellence, 2016), it is not yet understood if available services are acceptable, accessible, or if they improve outcomes as anticipated.

Conclusion

The literature on management of cognitive and behavioural changes in MND is sparse. The heterogeneity across research methods and outcome measures in the reviewed articles limits the strength of the existing evidence. Most peer-reviewed literature consists of expert commentary and there is a lack of primary data to guide HCPs and families on how to manage cognitive and behavioural change in MND. We propose that co-designed research in this space is needed to understand the preferences of pwMND and their families in

relation to the management of cognitive and behavioural changes. In addition, more standardised research practices to devise management methods or techniques for cognitive or behavioural impairment in MND are also needed. In line with the NICE (National Institute for Health and Care Excellence, 2016) guidelines, a randomised control study may determine if formal assessment for cognitive and behavioural deficits at diagnosis and/or repeated assessment improves clinical practice, subsequent care and quality of life for the pwMND and their carers.

Practice Implications

We propose that there is a clinical need to determine as early as practicable the presence of cognitive and behavioural changes in pwMND. This will enable HCPs to make adaptations to their approach to communication and to form partnerships with pwMND and their families. In turn, this will enable individualised care designed to support families to better understand, provide informed consent and enable care decisions to be made in partnership with families.

The previous chapter outlined current knowledge about management of cognitive and behavioural changes in MND. This information was collated from the existing evidence base and grey literature and highlights that most of the literature available to guide HCPs to manage cognitive and behavioural changes in MND, is derived from descriptive reviews or expert opinion. Additionally, the literature mainly contains general recommendations, with far fewer studies providing specific strategies or pathways for managing cognitive and behavioural changes in MND. Further, recommendations were drawn from research outside of the MND context e.g., within dementia research.

In the following two chapters, I explored how HCPs, working with families living with MND approach cognitive and behavioural changes in MND in their practice. Specifically, I aimed to understand how HCPs understand cognitive and behavioural changes, how they provide education about cognitive and behavioural changes to pwMND and carers, and how they manage cognitive and behavioural changes as part of their usual clinical practice. Chapter 6 describes a mixed methods study, aimed at understanding how HCPs manage cognitive and behavioural changes in MND. Specifically, this research project consisted of a convergent parallel mixed methods survey (Creswell, 2014; Moseholm & Fetters, 2017), which combined aspects of both quantitative and qualitative research (Creswell, 2008, 2014). Whilst surveys and mixed methods research are distinct methodological approaches, they can be applied in a single mixed methods survey study (Creswell & Hirose, 2019). It was important to seek the view of a broad range of HCPs because developing a better understanding of what clinical practice is occurring now to address cognitive and behavioural changes in MND will help to inform and design clinical pathways and future research to underpin better evidence-based practice for MND care.

CHAPTER 6: A SURVEY OF HCPS SUPPORTING PEOPLE WITH MND

Introduction

As outlined in Chapter 1, it is increasingly accepted that MND is a multisystem disease, with cell degeneration occurring beyond the motor cortices, resulting in impairments to functions other than purely motor control (Gallassi et al., 1985; MND Australia, 2020a; Neary et al., 2000b; Phukan et al., 2007). Cognitive and behavioural changes are now known to be part of the constellation of symptoms that are present in MND, said to occur in up to 50% of pwMND (Portet et al., 2001; Simon & Goldstein, 2019). These changes are reported to occur on a continuum with fronto-temporal dementia (FTD) (Motor Neurone Disease Association, 2022; Neary et al., 2000; Rusina et al., 2011), with up to 15% of people being diagnosed with MND/FTD (Motor Neurone Disease Association, 2022). Cognitive and behavioural changes in MND present as deficits in executive functioning, with apathy, frustration intolerance, awareness and insight, problem solving, learning and decision making often affected. Notably, evidence suggests that for some, non-motor symptoms may precede motor symptoms (Mioshi et al., 2014; Rusina et al., 2021).

Given the increasingly recognised prevalence of cognitive and behavioural changes in MND (Radakovic et al., 2024; Trucco et al., 2024), it is important to understand how these changes affect current clinical practices and symptom management. For example, it has been shown that in patients who present with cognitive and behavioural changes compared to pwMND who do not, treatment uptake may be reduced, health outcomes may be poorer, survival may be shorter and caregiver burden may be greater (Aoun et al., 2013; Chiò et al., 2012; N. H. Martin et al., 2014; Watermeyer et al., 2015). Given these significant detrimental impacts, developing a better understanding of the cognitive status of a pwMND, and how this might change across disease progression, would facilitate the implementation of timely and appropriate clinical management for both pwMND and carers. However currently, there is

little available evidence situated in MND-specific research to guide HCPs to manage cognitive and behavioural changes in MND (Francis et al., 2023).

Although MND clinical guidelines are available and include information about cognition decline as being associated with MND (National Institute for Health and Care Excellence, 2016), these guidelines are limited in that they provide general reference to a need to screen for, and manage, cognitive and behavioural changes. However, this information is derived from expert commentary and often drawn from dementia research. The current United Kingdom (UK) MND Clinical Guidelines and publications by the UK MND Association aimed at supporting health professionals, include recommendations to assess for, and regularly monitor for, cognitive and behavioural changes (Motor Neurone Disease Association, 2022; National Institute for Health and Care Excellence, 2016). These resources advocate for adaptations to care approaches in order to support clinical management of cognitive and behavioural changes. However, whilst HCPs working with pwMND may recognise that assessing for these changes in MND and subsequent management is important, the processes to do this appear unclear. For example, many HCPs may rely on clinical judgement informed by observation and carer report to identify cognitive changes (Crockford, Stockton and Abrahams, 2017). However, given the debilitating nature of MND, there is potential for cognitive and behavioural changes to be masked by other symptoms of the disease (Bak, 2010). For example, dysarthria is associated with MND, meaning the speech of a pwMND may become unintelligible, or they may lose the ability to speak completely (anarthria) due to the progressive decline in motor function. As such, a pwMND's inability to speak intelligibly, or at all, may limit a communication partner's ability to detect cognitive and behavioural changes that may occur. Similarly, pwMND experience impaired mobility often resulting in an inability to use their upper or lower limbs. Altered behaviours that may be observed when a person has the ability to move freely may not be detected, and behavioural changes may be missed. Given the insidious nature of cognitive and behavioural changes in MND, coupled with functional deficits, it is not known if cognitive

change in MND is overtly apparent, and is at risk of not being recognised or addressed clinically. Therefore, regular screening for, and assessment of, changes in cognitive function, appears critically important, and is addressed as such in MND clinical guidelines.

Current literature which addresses cognition in MND suggests that routine screening for cognitive or behavioural changes should occur early in disease and at regular intervals during disease progression (Burgos et al., 2018; National Institute for Health and Care Excellence, 2016). However, several barriers to implementing wide-spread routine cognitive screening have been identified (Crockford et al., 2017; Hodgins et al., 2018) and include resource deficits (specifically limited time and availability of staff to administer screening) as well as a lack of awareness of cognitive and behavioural changes, or a perceived unimportance to screen. Currently, clear pathways to guide HCPs to manage cognitive and behavioural changes, once identified are also lacking. Further, HCPs have reported their view that pwMND and their families may prefer not to know if cognition is affected. As such, this may result in HCPs not addressing cognitive and behavioural changes, in an attempt to alleviate further distress (Crockford et al., 2017).

In addition to a lack of clear diagnostic processes, there is also an acute lack of empirical data and evidence-informed resources to guide HCPs and families to manage cognitive and behavioural changes in MND (Francis et al., 2023). Resources that are available mostly consist of grey literature drawn from expert commentary, which is based on clinical experience or evidence derived from related areas outside of MND, e.g., dementia research. Whilst the literature previously identified is helpful, it consists of general recommendations and does not provide information related to the practices that are being enacted clinically, nor how the guidelines improve practice and patient outcomes.

It is important to understand the extent that cognitive and behavioural changes are being addressed in clinical practice and identify any barriers that exist to both assessing and managing cognitive and behavioural changes in MND, as this would help to inform and

design clinical pathways, professional development opportunities and future research to underpin better evidence-based practice in MND care. As such, it is critical to better understand how HCPs working with pwMND and their families currently receive training on, recognise, provide education for, and subsequently manage, cognitive and behavioural changes.

The research presented in this chapter outlines the findings of a survey of HCPs working with pwMND from various disciplines. The following research question was addressed:

How do HCPs recognise, provide information on, and manage, cognitive and behavioural changes in MND, and what enablers and barriers are there to this?

To support our understanding of the characteristics that may influence clinical implementation of management strategies for cognitive and behavioural changes in MND,and to help answer the research question, the Theoretical Domains Framework (TDF) (Cane et al., 2012) was used as a theoretical framework to support the interpretation of the findings. Applying theories of human behaviour and behaviour change provides opportunities to better understand the factors that influence practice and is a critical component of implementation science and behaviour change (Atkins et al., 2017). Using an approach, such as the TDF, helps to conceptualise complex issues, gives greater depth of interpretation, increases consistency, comparability, and relevance and aids in knowledge translation.

The TDF is underpinned by theoretical understanding of behaviour and behaviour change and the revised version consists of fourteen validated domains (Atkins et al., 2017; Cane et al., 2012). It allows for the systematic assessment of barriers and facilitators at the individual, team, and organisational level and in this study, will be used as an interpretive framework to identify potential barriers and enablers to implementing management of cognitive and behavioural changes in MND.

Methods

The aim of this research was to explore how HCPs who work with pwMND learn about, recognise, and provide education on cognitive and behavioural changes in MND. In addition, we aimed to better understand HCPs' attitudes, comfort with and approaches to, addressing, assessing and managing these changes. The TDF provided a framework to conceptualise how HCPs reported knowledge, skills and behaviours related to cognitive and behavioural changes in MND for their current practice, and informed opportunities for future practice.

We employed an online cross-sectional survey, as this approach allowed us to translate our research aims into measurable indicators and reach HCPs globally. A convergent parallel mixed methods study design (Creswell, 2014; Moseholm & Fetters, 2017) was chosen as this allowed for concurrent timing of data collection within a single phase of the research study, and for the questions to be designed to allow for simultaneous analysis of both the qualitative data and quantitative data. The survey aimed to canvas how HCPs from various disciplines who support families experiencing MND understand, recognise and manage cognitive and behavioural changes clinically.

Survey creation

Inclusion criteria

To be eligible to participate in this study, respondents needed to be a health/medical HCP currently working with, or having previously worked with, families experiencing MND. There was no limit to the years of practice or number of families with MND the HCP had supported. We included a range of HCPs from medical, allied health and nursing professions, as well as MND service providers and MND support coordinators. As this was an online survey, promoted via social media channels, HCPs working with MND in any location and from any area of practice were included. The survey was written in English.

Item generation

As part of the planning and design of this survey, we initially created survey items in a word document to clearly define concepts. Using an iterative approach, the broader research team discussed each survey question to reword, refine, and mitigate question bias (Blair et al., 2013). Items comprised of dichotomous (yes/no), multiple-choice, and matrix type questions relating to clinical practice and cognitive and behavioural changes in MND. These varied item types were included to reduce respondent time commitment, increase response rate and to assist with data interpretation. In addition, for specific questions which merited further explanation, the inclusion of an open-ended free text question followed directly after to allow respondents to elaborate on their answer (if they chose to do so). The item generation word document was used to obtain ethics approval (Appendix 5).

Item organisation

Once ethics approval was obtained from the Flinders University Human Research Ethics Committee (HREC #4660) (Appendix 1), the survey was created in the online survey platform, Qualtrics. The final iteration of the survey consisted of 55 items, which were approved by the ethics committee via an updated amendment. Question skip logic was applied within Qualtrics to create survey pathways to avoid respondents being asked irrelevant questions. The items were separated into the following content blocks:

- Block 1 consisted of a confirmatory question to meet the inclusion criteria. Respondents who answered “No” to the question “Do you work with people who have MND?” were automatically excluded from moving forward in the survey. The remaining items in this block included respondent characteristics and demographic questions (n=6/45, 13% of questions), including location, discipline, years of experience and an estimate of how many pwMND they had supported to date.
- Block 2 consisted of questions related to clinical practices specifically: providing education, identifying cognitive and behavioural changes, and management of cognitive and behavioural changes in MND. In addition, this section asked HCPs

about their level of comfort with discussing this topic with pwMND and their families (n=18/45, 40% of questions). Within question block 2, skip logic was applied to questions about cognitive and behavioural changes in MND. If a respondent answered, “Not a symptom of MND”, they were excluded from answering any further questions about cognitive and behavioural changes.

- Block 3 asked respondents about their personal experiences of receiving training and education about cognitive and behavioural changes in MND (n=6/45, 13%).
- Block 4 involved questions (n=13/45, 29%) related to clinical screening practices to identify cognitive and behavioural changes in MND. Respondents were asked questions related to the frequency of screening, the types of screening tools used, the benefits of conducting screening and the facilitators and barriers to conducting screening.

The final question in the survey invited respondents to voluntarily provide their contact details to participate in a follow-up interview where they could elaborate on the answers they provided in the survey. Respondents were advised that for participation in a follow-up interview they would receive an AUD$40 gift card honorarium. The results of these follow-up interviews are presented in Chapter 7.

Survey piloting

Prior to public survey dissemination, the survey was reviewed by staff members of MND South Australia (MND SA), who trialled the survey and provided feedback on the questions. Based on this feedback, the average completion time for the survey was approximately thirty minutes. The demographic section was amended, and the question of gender was removed as it was felt to be irrelevant. An ‘Other’ free text category was added to the question asking respondents about their discipline, to capture any area of practice that was not represented by the provided choices. As a result, the final survey contained 55 questions and 13 free text fields.

Data collection

Survey dissemination and recruitment

We aimed to recruit approximately 30 to 50 HCPs from any medical or health discipline or MND support service who work with families experiencing MND, globally. This sample size was considered feasible and realistic given the population of health care professionals who have experience working with an MND caseload, and the sample size is comparative to other studies (Anestis et. al, 2021). To achieve this, purposive and convenience sampling was conducted across a range of local and national interprofessional MND service providers. The survey link was disseminated via social media posts and through MND organisations within Australia, the UK and New Zealand. The MND Association in the UK helped to promote this research project within their professional community. The HCP call out was included in both the MND Association (UK) website and within their monthly newsletter to healthcare professionals. In addition, details of the project and the survey link were posted in the MND Association forum and via their social media channels. Further, the Australian Physiotherapy Association, Flinders University Caring Futures Institute, MND New Zealand, MND SA and MND Victoria included the survey link within their newsletters. [7]

In addition to the above recruitment strategy, a survey distribution list (Appendix 6) was created from publicly available email addresses relevant to any discipline involved in the care of persons with MND. This included but was not limited to private practices and various community organisations nationally and internationally. The dissemination email included a brief introduction to the survey aims and an anonymous survey link (Appendix 7). In addition, participants who expressed an interest in this research project were encouraged to invite additional eligible colleagues via a snowball recruitment strategy.

[7] Note: attempts to recruit via the ALS Association in the United States were unsuccessful. The Association advised that their support of research projects is limited to ALS Association grant funded studies.

The survey link was activated on the 20th of August 2022 and remained open until the 1st of March 2023.

Data preparation

At the end of the data collection period on the 1st of March 2023, there were a total of 78 responses. All Qualtrics survey data were individually examined within Qualtrics for pre-processing and to prepare for subsequent analysis. As this survey invited participants for a follow up interview and described an AUD $40 gift card, some respondents were identified as being potentially misrepresentative and consequently, their response required further examination. To do this, all questions, which included a free text answer were reviewed to identify if the text answer was relevant to the question. Further, the IP address of each respondent was viewed and any mismatch between the IP location and the respondent's location provided in their survey response was identified (e.g., the respondent picked the check box for Victoria, Australia (VIC), but the IP address was recorded in Miami, Florida). Finally, it was identified that all misrepresentative responses provided an email address which followed a consistent email address (Gmail) pattern. From this process, a total of 22 responses were identified as misrepresentative and removed. One additional response was removed as no survey questions were completed. Consequently, there were 55 responses included in data analysis.

To enable inductive analysis of the qualitative data (open ended questions that required a text response), all free text answers were extracted separately into an excel data file.

Data analysis

Quantitative analysis

Quantitative questionnaire data were analysed descriptively within Qualtrics using the inbuilt Results functionality. As this survey was exploratory in nature, descriptive analysis supported the identification of patterns and trends within the data. Additionally. given the size of the sample and the skew towards the speech pathology discipline, the research team

decided that descriptive data analysis was the most appropriate approach. Not all respondents completed each item of the questionnaire, therefore, where appropriate, the results are reported as a proportion of respondents per item.

Qualitative analysis

To describe characteristics of the responses, data from the qualitative questions (free-text responses) were analysed using an inductive content analysis approach (Kyngäs, 2020) to establish categories within these responses (Table 12). Free text answers for each question were compiled into a word document and reviewed. To identify patterns within these data, a process of conceptual analysis was conducted, considering word, sets of words and phrases relevant to each survey question. The research team iteratively reviewed these data to identify open codes. Following this, the open codes were combined with other codes with similar content to form categories.

The results from these qualitative and quantitative analyses are summarised below and presented in an integrated fashion to allow a descriptive rationale and deeper understanding of the meaning of the answers provided to the quantitative question.

Table 12.

Raw data coding process to domains of the Theoretical Domains Framework (TDF)

Survey question*: In your view, what prevents regular screening of cognitive and behavioural changes in MND in your role/service?*		
Raw data	Open coding	TDF Domain
Funding, complexity of client, caseload, *Funding available in NDIS space.*	Funding	
Raw Data	Open Coding	
Time available		
Time constraints	Time	
Nobody has the time to commit to doing it		Environmental Context and Resources
Raw Data	Open coding	
Staff		
Lack of specialist neuro psych support, (they can do it for those for those who are struggling with cognitive changes, but for all MND patients - currently - not resourced to screen and follow up everyone).	Lack of staff	
clinician availability		

Findings

Demographic characteristics

Respondents came from all states and territories of Australia, as well as the United Kingdom and New Zealand, representing 12 disciplines and six practice settings, as presented in Table 13. The research team are speech pathologists located in South Australia. This potentially explains the high number of respondents (n=19, 34.55%) that were speech pathologists in South Australia, which could also be further attributed to snowballing recruitment strategies. Despite the survey being disseminated via social media, no participants were recruited from American, Asian, African, European and Middle Eastern countries.

The length of experience working with pwMND varied, with the majority of respondents having between one and five years (n=19/55, 35%) or between five and ten years of clinical experience (n=16/55, 29%). Most respondents reported having supported between five to 20 pwMND (n=18/55, 33%) and 20 to 99 pwMND (n=21/55, 38%).

As it was not a requirement for respondents to provide an answer for every question in this survey, the overall respondent numbers changed per question. Therefore, the data are presented below to show the total number of respondents (from overall total of 55 respondents) who answered each question. Further, in a question that asked respondents to rank the importance of managing cognitive and behavioural changes in MND, one respondent selected ‘cognitive and behavioural changes are not applicable to MND’. Consequently, skip logic within the survey was applied and further questions regarding cognitive and behavioural change were not presented to this respondent.

Table 13.

Demographic characteristics of respondents

Characteristic		Count	Sample %
Location (n=55)	Australian Capital Territory	1	1.82%
	New South Wales	9	16.36%
	Queensland	7	12.73%
	South Australia	19	34.55%
	Tasmania	2	3.64%
	Victoria	5	9.09%
	Western Australia	1	1.82%
	New Zealand	3	5.45%
	United Kingdom	8	14.55%
Discipline (n=55)	Clinical care specialist	1	1.82%
	Dietician	1	1.82%
	Gastroenterologist	1	1.82%
	Neurologist	3	5.45%
	Occupational Therapist	6	10.91%
	Palliative Care	6	10.91%
	Physiotherapist	7	12.73%
	Psychologist/Neuropsychologist	2	3.64%
	Registered Nurse	5	9.09%
	Speech Pathologist	16	29.09%
	Support worker	1	1.82%
	Other	6	10.91%
Clinical Setting (n=55)	Specialty MND clinic	15	22.06%
	Private practice with MND caseload	8	11.76%
	Community organisation	28	41.18%
	Not for profit (NGO)	4	5.88%
	Residential aged care	2	2.94%
	Hospital	11	16.18%
	Less than 1yr	2	3.64%

Characteristic		Count	Sample %
Length of time (n=55)	1-5yrs	19	34.55%
	5-10yrs	16	29.09%
Length of time (n=55)	10-15yrs	10	18.18%
	20+yrs	8	14.55%
pwMND supported (n=55)	1-5	2	3.64%
	5-20	18	32.73%
	20-99	21	38.18%
	100+	14	25.45%

Describing clinical experience of cognitive & behavioural changes in MND

Ninety six percent (n=48/50) of respondents reported that they have observed clinical signs of cognitive and behavioural changes in pwMND. Thirty-seven of the 50 respondents (74%) provided a qualitative description of the changes they have observed. These terms are categorised as i. *Executive dysfunction, ii. Changes in behaviour and social cognition deficits, iii. Psychological descriptors, iv. FTD, v. Pseudobulbar affect and vi. Memory deficits.*

Table 14 represents the total number of instances respondents provided terms which were applicable to each category. Then within Tables 15, 16, and 17 a list of key words is shown with the frequency of use by respondents within each category.

Table 14.

Clinical signs of cognitive and behavioural changes in Motor Neurone Disease

Category	Frequency of term within each category
Executive dysfunction	46
Changes in behaviour and social cognition deficits	23
Psychological descriptors	26
Memory deficits	15
Pseudobulbar affect (emotional lability)	11
FTD	9

The most frequently provided terms in the category of *Executive dysfunction* were rigidity (11/46, 23%) and impaired language (9/46 19%) (Table 15).

Table 15.

Terms respondents used to describe executive function deficits in Motor Neurone Disease

Executive dysfunction (46)	Frequency of term
Rigidity	11
Impaired language	9
Impulsivity	6
Impaired decision-making	5
Impaired understanding	5
Lack of insight	4
Confusion	3
Impaired problem-solving	1
Impaired planning	1
Changes in executive function	1

The most frequently used terms in the category of *Behaviour changes and social cognition* were personality changes (7/23, 30%), however this was not further qualified in the survey responses, and reduced empathy or awareness of others (7/23, 30%) (Table 16).

Table 16.

Terms respondents used to describe behavioural changes in Motor Neurone Disease

Behaviour changes and social cognition (23)	Frequency of term
Personality changes	**7**
Reduced empathy or reduced awareness of others	7
Apathy	6
Reduced disinhibition	3

The most frequently used terms in the category of *Psychological descriptors* were frustration (4/26, 15%) and changes to mood (4/26, 15%) (Table 17). Anger and aggression were described by some respondents, however, were not further qualified in the survey responses.

Table 17.

Terms respondents used to describe psychological factors in MND

Psychological descriptors (26)	Frequency of term
Frustration	4
Changes to mood	4
Depression	3
Anger	3
Aggression	3
Hopelessness/Giving up/Despair	3
Anxiety	2
Agitation /Irritability	2
Suicidal ideation	1
Paranoia	1

Clinical discussions about cognitive and behavioural change in MND

When presented with a list of disciplines (Figure 20), respondents felt that it was primarily the responsibility of a neurologist or psychologist/neuropsychologist to discuss cognitive and

behavioural changes with families, with clinical care specialists and palliative care also suggested.

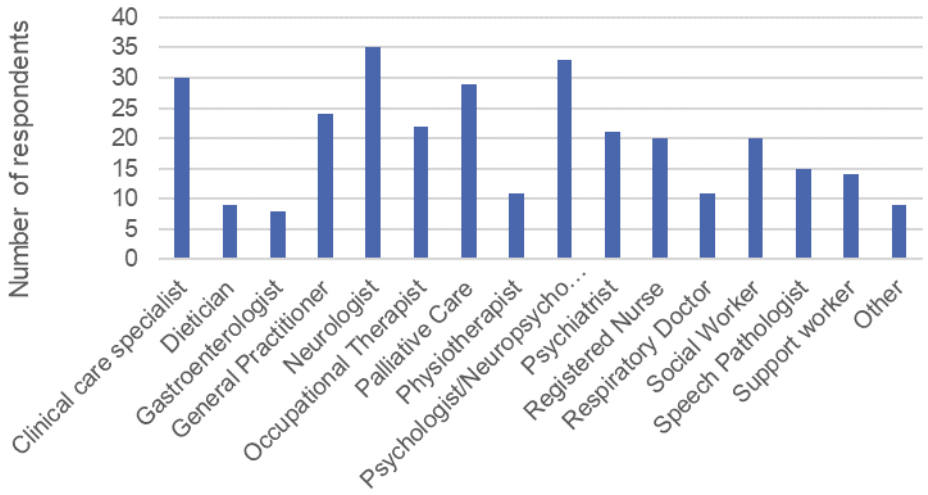


Figure 20: Survey question. Which discipline should discuss cognitive changes in MND? Respondents view of disciplines responsible for discussing cognitive and behavioural changes with families.
(MND: Motor Neurone Disease). **Image created by thesis author.**

When asked about levels of their own comfort related to discussing cognitive and behavioural changes with pwMND (Figure 21), the majority of respondents felt either somewhat comfortable (n=18/39, 46%) or extremely comfortable (n=12/39, 31%) having these conversations with pwMND. This was similarly reflected in conversations with families (Figure 20).

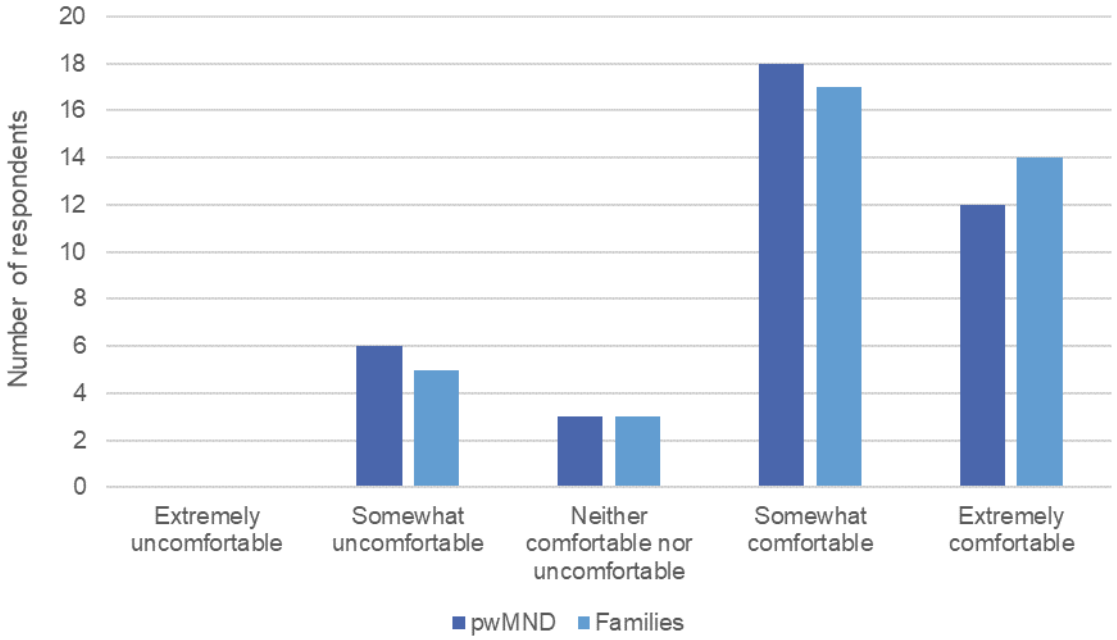


Figure 21: Survey question. HCPs' level of comfort discussing cognitive and behavioural change with pwMND and their families.
(HCPs: Health care professionals; pwMND: people with Motor Neurone Disease)

When asked to express their view on whether they thought that pwMND would want to know about cognitive and behavioural changes associated with MND, respondents were evenly split (*n*= 19/39, 49%). Respondents who expressed that pwMND did not want to know about cognitive and behavioural changes associated with MND described prioritising the visible symptoms of the disease and dealing with the challenges of the physical changes.

> Their focus tends to be on extending their life expectancy, remaining mobile, remaining able to interact with their loved ones *(Survey respondent 63).*

Respondents whose view it was that pwMND do want to know about cognitive and behavioural changes suggested this was to better understand the disease, feel prepared and feel in control.

> I think a lot of MND patients want to know what's 'ahead' of them and what they/their families can expect. So, when things happen or change, they may be a little more prepared than otherwise (Survey respondent 10).

Additionally, respondents all described that in MND each case is different, and there is not a one size fits all approach to this question.

> As with all things MND there is no one answer that would suit all MND patients. Some patients want to know absolutely everything. Others just take one day at a time and do not want to know what may/may not be ahead (Survey respondent 14).

In contrast, respondents perceived that the majority of carers of pwMND wanted to know about cognitive and behavioural changes (n= 34/38, 89%) with only four (n= 4/38, 11%), suggesting carers do not want to know. Respondents provided reasoning for their views in the qualitative data, suggesting that carers want to know about cognitive and behavioural changes to better support their understanding of the disease, to feel prepared, to manage behavioural changes and to better support their loved one.

> …understand that these changes are being caused by disease progression and not a true reflection of the person (Survey respondent 11).

> Families most definitely wish to understand and know more about the reasons why their loved one may have changed in subtle ways. They seek to be heard and validated with the concerns and are often comforted by the news that this can be part of the disease process (Survey respondent 68).

Respondents, whose view it was that carers do not want to know about cognitive and behavioural changes, described emotional stress and acceptance as the reasons.

> Most families just try to ignore any changes. (Survey respondent 04).

> The families I have worked with are often experiencing grief and loss differently, and in many ways, grieve the loss to their own lives i.e., providing additional care to a loved one who was previously fully independent on top of their current workload. I find that family members find it more difficult to process the changes a pwMND is likely to experience and tend to focus on the physical and communication changes. (Survey respondent 13).

As shown (Figure 22), the majority of respondents reported discussing cognitive and behavioural changes in MND with families most of the time (n=17/39, 44%) or always (n=3/39, 7%). Other respondents described that this was not part of their routine practice, stating they discussed this either half of the time (n=9/39, 23%) or sometimes (n=8/39, 21%). Two respondents (5%) described that they never discuss cognitive and behavioural changes with families.

Most respondents reported that families experiencing MND sometimes asked them about cognitive and behavioural changes (n=20/39, 51%) (see Figure 23). The remainder suggested that they are never asked (n=10/39, 26%), asked half of the time (n=5/39. 13%) or asked most of the time (n=4/39, 10%). No respondent described being asked by families about cognitive and behavioural changes all the time.

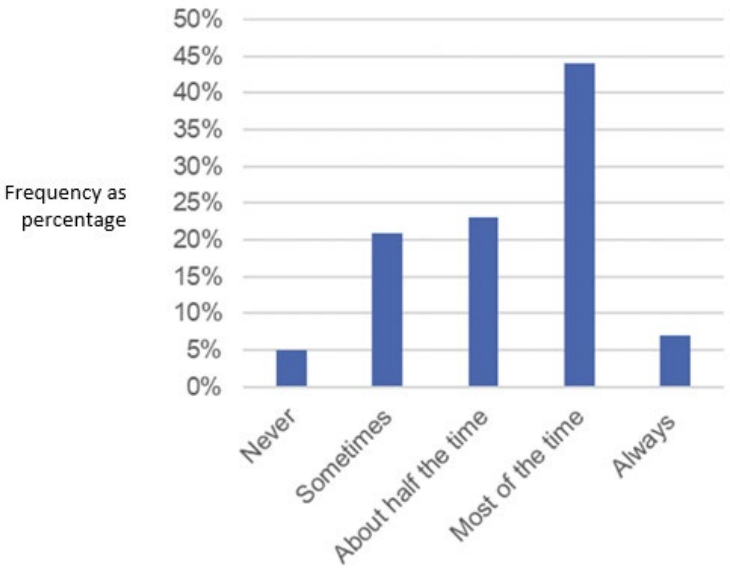


Figure 22: Survey question - How often do you discuss cognitive and behavioural changes in MND with families? (n=39/55) respondents.
(MND: Motor Neurone Disease)

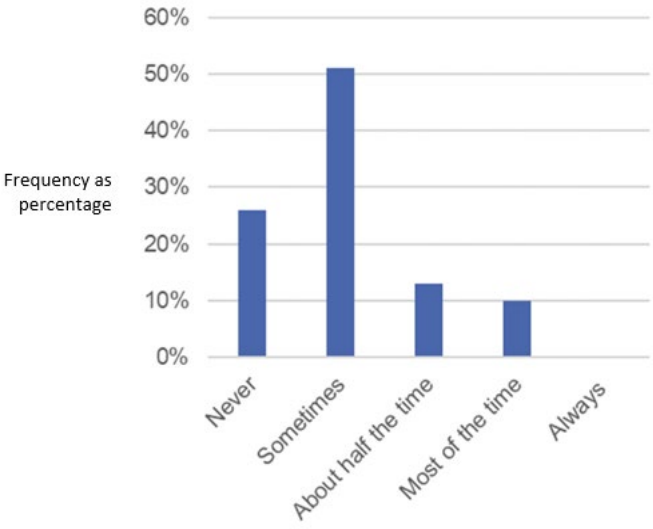


Figure 23: Survey question - How often are you asked about cognitive and behavioural changes in MND by families? (n=39/55) respondents.
(MND: Motor Neurone Disease)

Accessing information on cognitive and behavioural change in MND

Thirty-four (n=34/55, 62%) respondents described ways in which they accessed information about cognitive and behavioural change in MND, with the majority using publicly available MND specific services (n=19/34, 56%) or peer-reviewed literature (n=14/34, 41%), while others described discussion with colleagues (n=8/34, 23%), attendance at professional development courses or workshops (n=9/34, 26%), or reliance on their own clinical experience (n=3/34, 8%). A small number of respondents described unspecified websites (n=9/34, 26%), books (n=2/34, 5%), and podcasts (n=1/34, 2%).

In addition, respondents (n=15/34, 44%) provided weblinks to the documents that they source to inform them about cognitive and behavioural changes in MND and a small number of respondents (n=5/34, 15%) uploaded these resources. A summary of these resources is shown in Table 18.

Table 18.

Web links and documents provided by health care professionals regarding cognitive and behavioural changes in Motor Neurone Disease

Web address	Website name	Country	Count
ttps://www.mndaustralia.org.au/	MND Australia	Aust	1
https://mnd.org.nz/	MND New Zealand	NZ	1
https://www.mndandme.com.au/	MND and Me Foundation	QLD, Aust	1
https://www.nice.org.uk/guidance/ng42	NICE guideline [NG42}. Motor neurone disease: assessment and management	UK	1
https://mndnsw.org.au/information-professionals.html	MND NSW Information for health and community care professionals	NSW, Aust	1
https://www.mndassociation.org/	MND Association	UK	1
https://www.mndaustralia.org.au/mnd-connect/living-with-mnd/cognition-thinking-behaviour	MND Connect. Cognition, thinking and behaviour	Aust	
https://www.mndassociation.org/app/uploads/2020/07/Changes-to-thinking-and-behaviour-with-MND.pdf	MNDA. Emotions, thinking and behaviour	UK	3
https://www.mndaustralia.org.au/mnd-connect/information-resources/cognitive-and-behaviour-change-in-mnd	MND Connect. Factsheet. Cognitive and behaviour change in MND	Aust	3
https://www.mndaustralia.org.au/mnd-connect	MND Connect. Information and support.	Aust	1
https://www.mndassociation.org/support-and-information/	MNDA. Support and information	UK	1

Web address	Website name	Country	Count
https://www.mndassociation.org/app/uploads/px018-cognitive-change-ftd-mnd.pdf	Not found	UK	1
https://healthtalk.org/motor-neurone-disease-mnd/thinking-and-behaviour-with-mnd	Healthtalk.org. Thinking and behaviour with MND	UK	1
Source unknown (Created by MND physician March 2011)	Cognitive changes in MND.doc	Unknown	1
https://www.mndaustralia.org.au/mnd-connect/information-resources/cognitive-and-behaviour-change-in-mnd	MND Factsheet_ Cognitive & Behaviour Change _ MND Australia _ MND Australia.pdf	Aust	2
Living Better for Longer: MND Australia Fact Sheet EB2 (2014) (www.mndaust.asn.au)	Cognitive-and-behaviour-change.pdf	Aust	1
https://www.mndassociation.org/sites/default/files/2022-11/Changes%20to%20thinking%20and%20behaviour%20with%20MND.pd	Changes-to-thinking-and-behaviour-with-MND.pdf	UK	1

Screening for cognitive and behavioural changes in MND

Initial contact screening and follow-up screening

Of the 36 respondents who answered questions regarding screening practices, nearly half (n=17/36, 47%) reported that screening (as routine clinical practice) was never completed in the initial contact with a person with confirmed or probable MND (Figure 24). Respondents were also asked about screening practices for other neurodegenerative disease. Nearly half also reported that screening (as routine clinical practice) was never completed in the initial contact with a person for other neurodegenerative conditions (n=16/34, 47%).

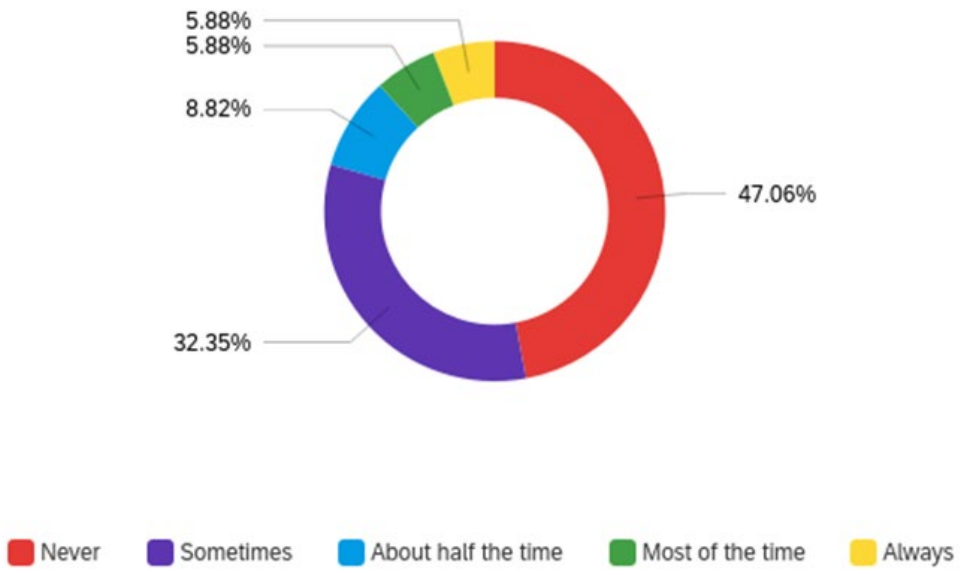


Figure 24: Respondents describing clinical screening practices for cognitive and behavioural changes in MND.
(MND: Motor Neurone Disease)

Respondents reported to conducting follow-up screening 35% of the time (n=7/20), however, 65% (n=13/20) of respondents reported that regular screening was not conducted as part of routine practice.

Screening/assessment tools

The Montreal Cognitive Assessment (MoCA) and Mini-mental state examination (MMSE) were the most frequently selected cognitive assessments by respondents (n=7/36, 19%) with the Frontal Assessment Battery (FAB) and the Edinburgh Cognitive and Behavioural ALS Screen (ECAS) also prominently reported (n=6/36, 17%) (Figure 25 shows all screenings tools used).

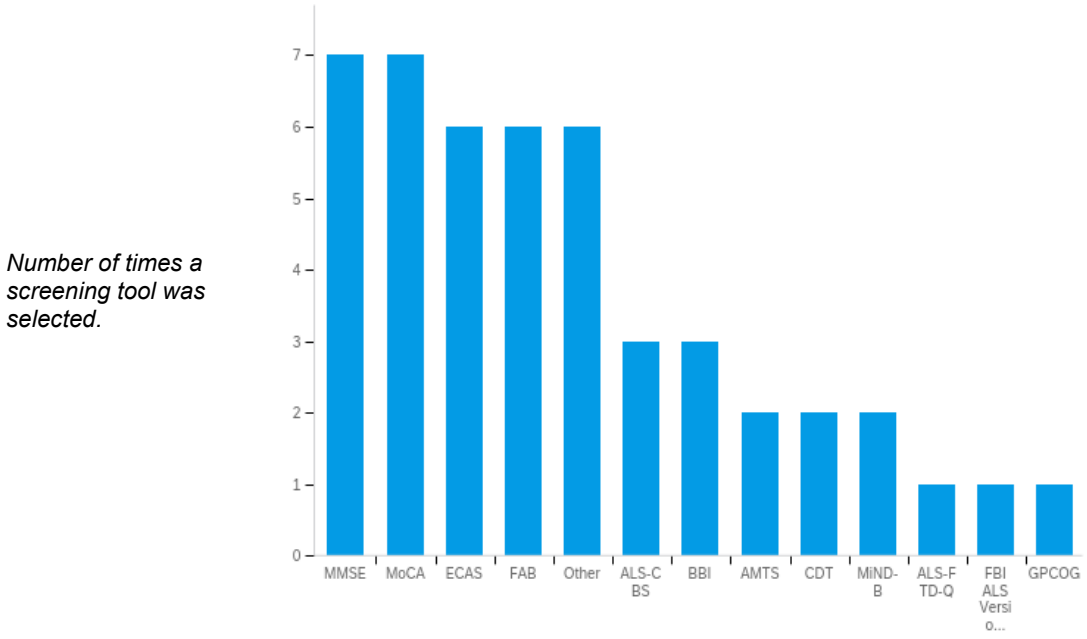


Figure 25: Screening tools for cognitive and behavioural changes in MND described by respondents.
(MND: Motor Neurone Disease; Screening tools for cognitive and behavioural change in MND described by respondents. MMSE = mini-mental state exam, MoCA = Montreal, ECAS = Edinburgh cognitive assessment screen, FAB = frontal assessment battery, ALC-CBS = amyotrophic lateral sclerosis cognitive and behavioural screen, BBI =Beaumont Behavioural Interview , AMTS = Abbreviated Mental Test score , CDT =Clock Drawing Test , MiND-B =Motor Neuron Disease Behavioural Scale , ALS-FTD-Q = ALS Frontotemporal Dementia Questionnaire, FBI ALS =Frontal Behavioural Inventory ALS Version , CPCOG = General Practitioner Assessment of Cognition)

Fifty percent of respondents (n=18/36) felt that screening for cognitive and behavioural changes in MND was moderately important (Figure 26), with 25% (n=9/36) reporting that they felt it was very important and 13% (n=5/36) felt that it was extremely important. Eleven percent of respondents (n=4/36) felt that screening for cognitive and behavioural changes was slightly important. None of the respondents of this question selected the option of 'not at all important'.

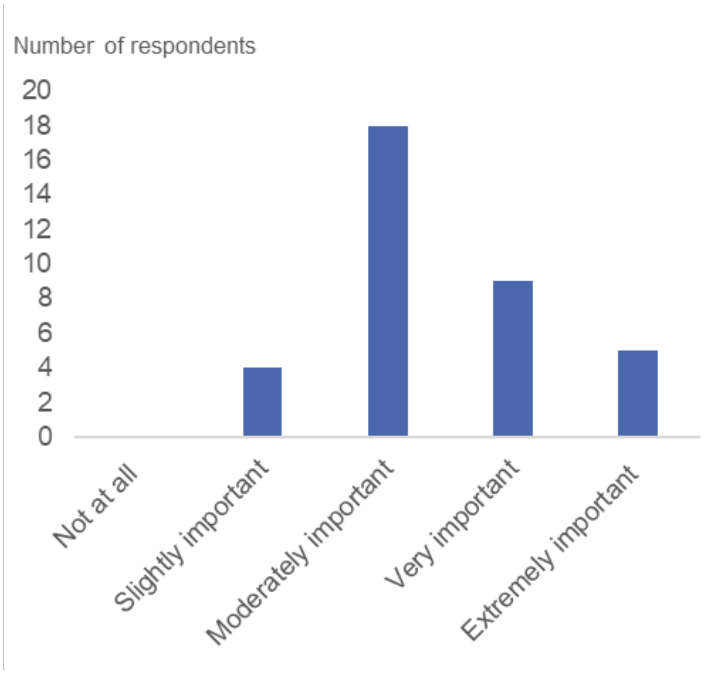


Figure 26: Survey question - How important is screening for cognitive and behavioural changes in MND? (n=36/55 respondents)
(MND: Motor Neurone Disease)

Of the 36 respondents, 31 (86%) elaborated on their response. Respondents identified screening allowed for a baseline measurement of cognitive and behaviour, and facilitating conversations about advanced care planning, capacity and to direct management. Respondents also identified that routine screening helps to normalise cognitive and behavioural changes that may be associated with MND for some.

> Routine screening provides an opportunity to normalise the chance of cognitive/behavioural change, to identify potential problems early (sometimes) and to open up conversations with family members/carers and staff… (Survey respondent 41)

In contrast, other respondents reported that screening was not conducted as part of routine practice and felt that screening may create additional burden for pwMND and their families.

> The pwMND or their family may not be focussing on cognitive at that point, it may be too distressing usually only screen/review if they report something themselves. (Survey respondent 04)

Of the 55 respondents, thirty-seven (67%) identified several facilitators for cognitive and behaviour screening. The most prominent categories for faciliating screening included multidisciplary collaboration (n=9/37, 24%), skilled staff and availability of resources (n=8/37, 22%), and ongoing and regular client follow up (n=7/37, 19%).

> A dedicated team who manages this patient group. Collaboration between all parties of an MDT (Survey respondent 68)

> Having time and resources (suitably trained staff, with access to neuropsychology) to complete screening in a meaningful way. (Survey respondent 41)

> Regular visits/assessments and reviews, to better understand cognitive changes as they appear. (Survey respondent 11)

In addition, family report of cognitive changes was prominently identified as a facilitator to screening (n=6/37, 16%). The less prominent facilitators to screening for cognitive change included pwMND self-reporting symptoms (n=2/37, 5%), developing a strong relationship with families living with MND (n=2/37, 5%) and having a clear clinical pathway (n=1/37, 2%).

Of the 55 survey respondents, 30 (n=30/55, 55%) answered the question asking their view on what were barriers to cognitive and behavioural screening in MND. The most prominent barrier identified by respondents was a lack of resources (n=24/30, 80%), specifically related to insufficient time (n=12/30, 40%), insufficient skill (n=6/30, 20%), insufficient staffing (n=5/30, 17%) or insufficient funding (n=2/30, 6%).

> Funding, time available, complexity of client, caseload, HCP availability (Survey respondent 02)

> Lack of specialist neuro psych support, (they can do it for those for those who are struggling with cognitive changes, but for all MND patients - currently - not resourced to screen and follow up everyone (Survey respondent 53)

> Nobody takes ownership of it. Most likely because nobody has the time to commit to doing it (Survey respondent 52)

Respondents futher suggested that prioritising other symptoms of MND (n=6/30, 20%), a lack of access to service/client (n=4/30, 13%), barriers within the service model design (n=6/30, 20%), relationship with families (n=7/30, 23%) or unclear benefits to conducting screening (n=2/30, 6%) were also barriers to regular screening practices for cogntive and behavioural change in pwMND.

> …the client competing priorities and needs take priority of completing MND cognitive assessments. (Survey respondent 01)

> Time, there are sometimes more important things to discuss in the clinic. (Survey respondent 71)

> Rapid deterioration when trying to push forward life extending treatments such as NIV and PEG. Lack of specialist neuro, psych support. Not resourced to screen and follow up everyone (Survey respondent 53)

> Lack of rapport with the person and their family. Lack of follow through as this impacts on trust. (Survey respondent 13)

Identifying cognitive and behavioural changes when communication is impaired

Thirty-nine respondents (71%) elaborated on the methods they used to identify cognitive and behavioural changes in pwMND who present with impaired speech (dysarthria/anarthria), with the following categories described. Clinical (indirect) observation (n=20/39, 51%) and carer report (n=19/39, 49%) were the most frequently described methods for detecting cognitive and behavioural changes in the presence of dysarthria. Informal assessment methods (n=8/39, 21%) and history taking (n=6/39, 15%) were also described, with fewer describing undertaking a formal assessment (n=4/39, 10%).

Within this question, respondents also identified using alternative and augmented communication (AAC) methods. These consisted of both high-tech AAC e.g., electronic devices and low-tech AAC e.g., non-electrical methods to support their assessment for cognitive and behavioural changes, if the person had functional communication difficulties related to dysarthria (n=14/39, 36%).

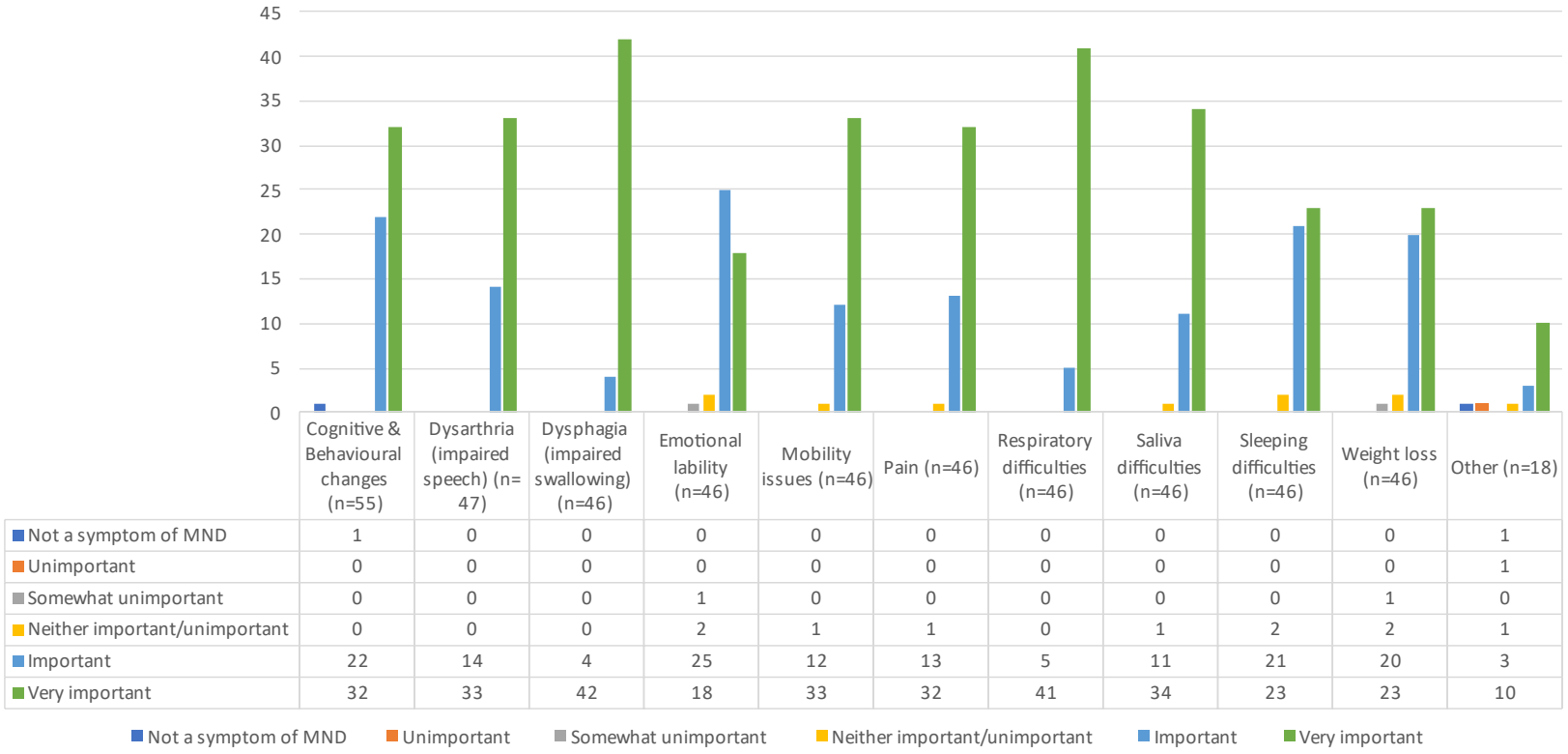
Some respondents described that cognitive and behavioural changes may be difficult to detect (n=1/39, 2%) or that these changes may not be detected at all (n=2/39, 5%). Referral to another discipline was also described (n=4/39, 10%).

Clinical management of cognitive and behavioural changes in MND

Respondents were asked to rank the level of importance of managing common MND symptoms from unimportant to very important (Figure 27). Of these, 98% of respondents (n=54/55) ranked cognitive and behavioural changes as important or very important (see Figure 26). Cognitive and behavioural changes were ranked as having similar importance to the more commonly recognised symptoms of MND being dysphagia (n=46/46, 100%), respiratory difficulties (n=46/46, 100%), mobility issues (n=45/46, 98%) and pain (n=45/46, 98%). [8]

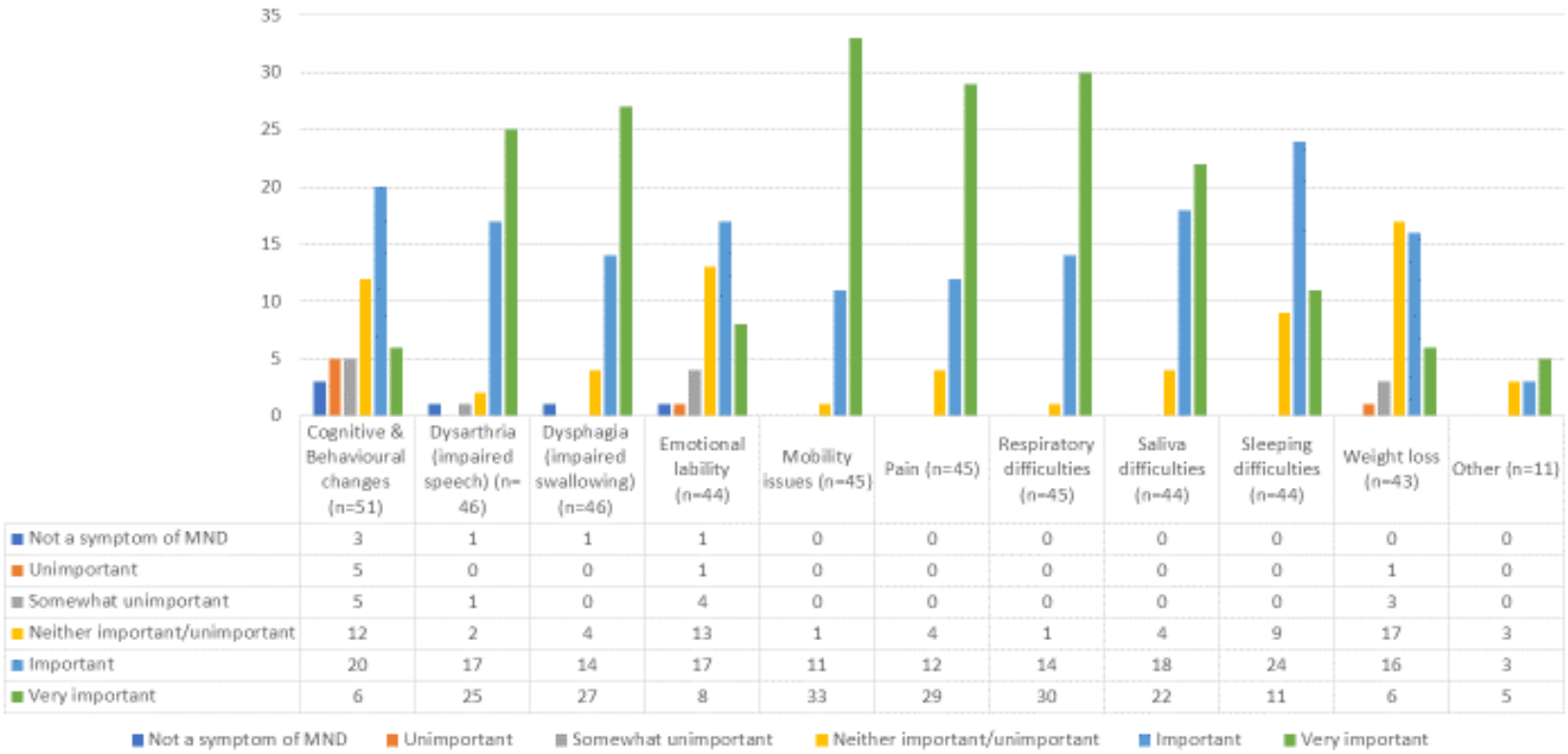
When respondents were asked their view about which MND symptoms people with the disease felt were the most important to manage, the results were more varied. Respondents suggested that pwMND viewed mobility issues (n=44/45, 98%) and respiratory difficulties (n=44/45, 98%) as the most important to manage, with dysarthria (n=42/46, 91%) dysphagia (n=42/46, 91%) also ranking highly (Figure 28). Respondents expressed their belief that pwMND viewed management of cognitive and behavioural changes as important (n=26/51, 51%), neither important or unimportant (n=12/51, 24%) and five respondents (9%) believed that this symptom was unimportant to pwMND to manage.

[8] Note: One HCP responded that cognitive and behavioural changes were not applicable to the disease. Skip logic within the survey was applied to this question and further questions regarding cognitive and behavioural change were consequently not presented to this respondent.



	Cognitive & Behavioural changes (n=55)	Dysarthria (impaired speech) (n=47)	Dysphagia (impaired swallowing) (n=46)	Emotional lability (n=46)	Mobility issues (n=46)	Pain (n=46)	Respiratory difficulties (n=46)	Saliva difficulties (n=46)	Sleeping difficulties (n=46)	Weight loss (n=46)	Other (n=18)
Not a symptom of MND	1	0	0	0	0	0	0	0	0	0	1
Unimportant	0	0	0	0	0	0	0	0	0	0	1
Somewhat unimportant	0	0	0	1	0	0	0	0	0	1	0
Neither important/unimportant	0	0	0	2	1	1	0	1	2	2	1
Important	22	14	4	25	12	13	5	11	21	20	3
Very important	32	33	42	18	33	32	41	34	23	23	10

Figure 27: Survey question - HCPs' view of what symptoms of MND are important to manage.
(HCPs: Health care professionals; MND: Motor Neurone Disease)



	Cognitive & Behavioural changes (n=51)	Dysarthria (impaired speech) (n=46)	Dysphagia (impaired swallowing) (n=46)	Emotional lability (n=44)	Mobility issues (n=45)	Pain (n=45)	Respiratory difficulties (n=45)	Saliva difficulties (n=44)	Sleeping difficulties (n=44)	Weight loss (n=43)	Other (n=11)
Not a symptom of MND	3	1	1	1	0	0	0	0	0	0	0
Unimportant	5	0	0	1	0	0	0	0	0	1	0
Somewhat unimportant	5	1	0	4	0	0	0	0	0	3	0
Neither important/unimportant	12	2	4	13	1	4	1	4	9	17	3
Important	20	17	14	17	11	12	14	18	24	16	3
Very important	6	25	27	8	33	29	30	22	11	6	5

Figure 28: Survey question - HCPs' view of what pwMND feel are important symptoms of MND to manage.
(HCPs: Health care professionals; MND: Motor Neurone Disease)

Thirty-nine respondents (n=39/55, 71%) provided a description of their clinical approach to managing cognitive and behavioural changes associated with MND, including a multi-disciplinary approach (n=11/39, 28%) with referral to their wider clinical team (n=10/39, 26%) and providing support to the carer (n=10/39, 26%). Education about cognitive and behavioural changes in MND was described by 36% of respondents (n=14/39), with the recipient of this education being the pwMND (n=4/14, 29%) or the carer (n=4/14, 29%), specifically. Six of the 39 respondents (15%) did not specify who would be the recipient of the education. Adaptations to communication were described as a management strategy by 18% of respondents (n=7/39). Finally, 23% (n=9/39) of respondents suggested that medications, such as antidepressants, would be a management strategy.

In summary, respondents felt cognitive and behavioural changes in MND are important to manage, however, approximately half of respondents perceived that pwMND did not identify cognitive and behavioural changes as important to manage, whilst the remainder identified this as important for pwMND. Respondents expressed their view that pwMND felt mobility and respiratory impairments were the most important symptoms to manage. The management strategies described by respondents included MDT involvement, referrals to another discipline, supporting the carer, providing education and adapting communication. Medical intervention (antidepressants) was also described as a management strategy for cognitive and behavioural changes.

HCPs training to manage cognitive and behavioural changes in patients

Of the 39 respondents, 22 (56%) reported that they had not received specific training to manage cognitive and behavioural changes in people with neurodegenerative conditions in general, and 28 (72%) described not having received training to manage cognitive and behavioural changes specific to MND. Of those respondents who received previous training (n=9/39 23%), the majority reported this was conducted by a different discipline to their own with neuropsychologist (n=2/9, 22%), occupational therapist (n=1/9, 11%), palliative care

(n=1/9, 11%) and geriatrician (n=1/9, 11%) described. Training from previous University studies (n=2/9, 22%), as well as training provided by MND associations (n=2/9, 22%) was also reported. One respondent described learning on the job (n=1/9, 11%).

In summary, respondents mostly described that they have not received specific training to manage cognitive and behavioural changes in MND or in other neurodegenerative disease.

Summary of findings

The following table summarises the findings of this study. The TDF domains of *Knowledge*, *Belief about consequences* and *Belief about capabilities* are facilitators to managing cognitive and behavioural changes in MND. Whereas the TDF domains of *Skills*, *Social professional role* and *Environmental context and resources* and identified as barriers to managing cognitive and behavioural changes in MND (Table 19)

Table 19.

Facilitators and barriers to clinical management of cognitive and behavioural changes in Motor Neurone Disease within the Theoretical Domains Framework domains

TDF Domain	Facilitators	Barriers
Knowledge	*HCPs know that cognitive and behavioural changes are associated with MND.*	*Evidence regarding practice for cognitive and behavioural changes is under-developed*
Skills		*Training to manage cognitive and behavioural changes in MND is lacking* *Limited skilled staff*
Social professional role		*Unclear whose responsibility it is to address and manage cognitive and behavioural changes*
Environmental Context and Resources		*Limited resources* *Prioritisation of practice that relates to immediate acute concerns* *Limited access to neurology services*
Belief about consequences	*HCPs believe managing cognitive and behavioural changes in MND is important*	*HCPs were divided in their belief if pwMND wanted to know about cognitive and behavioural changes* *HCPs described this information may overwhelm or upset people*
Belief about capabilities	*HCPs are comfortable discussing cognitive and behavioural changes with families*	

Discussion

We explored current clinical practices related to cognitive and behavioural changes in MND via an online cross-sectional survey. To support the interpretation of these data, we applied the TDF (Atkins et al., 2017; Cane et al., 2012) to assist in understanding the facilitators and barriers to addressing and managing cognitive and behavioural changes in MND.

The facilitators to managing cognitive and behavioural changes in MND are summarised as: i. HCPs know that cognitive and behavioural changes are associated with MND, ii. HCPs believe cognitive and behavioural changes are important to manage, and iii. HCPs are comfortable in their capability to discuss cognitive and behavioural changes. Additionally, the barriers to managing cognitive and behavioural changes in MND are summarised as: i. training to manage cognitive and behavioural changes in MND is lacking and there are limited skilled staff, ii. there are resource barriers and limited access to neurology services, iii. it is not clear which HCPs are responsible for managing cognitive and behavioural changes. These themes will be discussed within the domains of the TDF framework.

The TDF domain of *Knowledge* may be defined as an ‘awareness that something exists’ and includes the construct of ‘knowledge of condition’ (Cane et al., 2012). For this study, the domain of *Knowledge* was interpreted as the knowledge construct that cognitive and behavioural changes exist in MND. These findings show that knowledge was a facilitator in addressing and managing cognitive and behavioural changes in MND. Specifically, most respondents know that cognitive and behavioural changes are associated with MND, with most reporting they have observed these changes in their clinical practice. Further, respondents described characteristics of cognitive and behavioural changes that are mostly consistent with the MND literature, with executive functioning deficits prominently described (Tjokrowijoto et al., 2023).

Deficits in executive functioning are important to consider in clinical management of MND, as these deficits have been shown to negatively impact on an individual’s ability to understand

complex information and make decisions about their ongoing care (White et al., 2023). In addition to impaired executive functions, pwMND may also develop language deficits, which can impact on word finding and verbal fluency, and be associated with difficulties expressing thoughts and feelings (Neary et al., 2000). Additionally, social cognition may be impaired in some pwMND, with reduced theory of mind, reduced empathy, reduced social perception and abnormal social behaviours described (Henry et al., 2016; Lillo et al., 2020; Tjokrowijoto et al., 2023). Consequently, it is important for HCPs to determine a pwMND's cognitive status, as both executive function deficits and social cognition deficits may be mistaken as reduced mood (depression), inappropriate responses to questions, a lack of interest or initiative or a perceived lack of adherence to management strategies (Neary et al., 2000). Within this study, HCPs additionally noted the psychological descriptors of anxiety and depression as being a characteristic of cognitive and behavioural changes in MND. However, it is unclear if these observations are due to a pwMND's psychological response to a terminal disease diagnosis, or if these presentations are associated with neurological changes that occur along the MND FTD continuum.

Differentiating between neurological changes and a normal psychological/grief response in MND is important to guide management. Notably, antidepressant medication was described by 23% of respondents as a management strategy for cognitive and behavioural changes in MND. Whilst selective serotonin reuptake inhibitors (SSRIs) are reported to be effective to manage depression and pseudobulbar affect, i.e., psychological responses to MND, the literature is unclear regarding the effectiveness of SSRIs for managing cognitive and behavioural changes (Arora & Khan, 2020; Miller et al., 2009; Shoesmith et al., 2020; Thompson, 2019). Some characteristics of cognitive and behavioural changes in MND, such as inhibition and impulsivity, are reported to improve with SSRIs (Arora & Khan, 2020). However, currently there is insufficient evidence that SSRIs are effective as a management strategy for cognitive and behavioural changes in MND (Arora & Khan, 2020). An added layer of complexity found in this study, which presents as an additional barrier to addressing

and managing cognitive and behavioural changes, is related to the respondent's professional development.

The constructs of 'skill development' and 'competence' are situated within the TDF Domain of *Skills*. Respondents reported that they had not received training for cognitive and behavioural changes in MND and most had not received training for cognitive and behavioural changes in any other type of neurodegenerative disease. Similarly, inadequate staff training has been found to be a barrier for conducting cognitive screening in an MND population (Crockford et al., 2017). Subsequently, HCPs may feel unsure about how to approach cognitive and behavioural changes in MND. This lack of training may be one possible explanation for respondents describing making referrals to a different discipline as a way to manage cognitive and behavioural changes.

The TDF Domain of *Social/Professional role and Identity* includes the construct of 'professional roles'. Respondents were not clear on who is responsible for managing cognitive and behavioural changes in MND. Whilst neurology and palliative care were the most frequently identified disciplines, respondents also selected multiple other disciplines. This may be considered as both a facilitator and a barrier to addressing cognitive and behavioural changes in MND. Namely, the wide range of disciplines selected may mean that respondents believe that each person within an MDT has an active role in supporting families with cognitive and behavioural changes. Conversely, it may also be seen that currently it is not clear who is responsible for addressing and managing cognitive and behavioural changes, reflecting the risk that no one will take on this responsibility. This lack of clarity is similarly reflected in the publicly available MND information, which is designed to support pwMND experiencing concerns about cognitive and behavioural changes. For example, some information suggests contacting a "health professional" (MND Australia, 2020a), a GP, or a "relevant professional' for concerns about cognitive and behavioural changes. However, the complexity of the generality of this information is three-fold. First,

stigma has been shown to be associated with cognitive changes, both in society and in health care (Motor Neurone Disease Association, 2022; Warren, 2023), and cognitive changes are frequently not reported by people. Second, a pwMND may lack the insight and awareness that these changes are occurring (Motor Neurone Disease Association, 2022). Third, families may not be aware that cognitive and behavioural changes in MND can occur and may not seek out the information. Considering these factors, it could be presumed that the responsibility for addressing cognitive and behavioural changes lies with the families. However, this is not ideal, and more needs to be done to both train HCPs and develop clearer clinical pathways to address ownership of the management of this symptom of MND. However, when this is considered in the TDF domain of *Environmental Context and Resources,* further complexities are identified.

Environmental Context and Resources includes the constructs of 'environmental stressors', 'resource availability' and 'person x environment interaction'. Collectively, all respondents reported that both staff training opportunities and access to resources in their current clinical model as environmental barriers. The lack of resources to support HCPs to manage cognitive and behavioural changes is similarly reflected in Chapter 5 (Francis et al., 2023). This review found that resources, which describe strategies to manage cognitive and behavioural changes in MND, are limited and mainly consist of general information based on expert commentary. Further, these environmental challenges have been shown to be an explicit barrier to conducting screening for cognitive and behavioural changes in MND (Crockford et al., 2017). Combining these factors, namely i. an absence of professional development opportunities, ii. a potential lack of discipline specific ownership, and iii. lack of resources, there is a risk that cognitive and behavioural changes in MND may be overlooked. Furthermore, it is not clear from the findings if respondents believed if it was beneficial to discuss cognitive and behavioural changes with families living with MND.

The TDF domain of *Belief about consequences* featured as both a facilitator and barrier to addressing and managing cognitive and behavioural changes in MND. First, some respondents believed that cognitive and behavioural changes in MND are an important symptom to manage. This facilitating factor presents as an opportunity to develop strategies that support HCPs to implement cognitive and behavioural change conversations with families as part of routine practice. However, as demonstrated in the behaviour change literature (Atkins et al., 2017; Cane et al., 2012), it is important to also identify a learning model which considers a HCPs' potential to acquire knowledge about cognitive and behavioural changes in MND as well as determining both self-efficiency and motivation to change behaviours in clinical practice.

Interestingly, respondents were divided in their belief if pwMND would not want to know about cognitive and behavioural changes in MND or not. Some believed that pwMND were already overwhelmed from receiving a terminal diagnosis and experiencing the physical challenges of MND. Subsequently, a barrier to discussions about cognitive and behavioural changes as associated with MND are related to the respondents' beliefs that these conversations would have a negative psychological consequence. Some respondents expressed their belief that bringing up cognitive and behavioural changes may create relational barriers and break trust between the HCP and their patients. A similar finding has also been described as a barrier to implementing cognitive screening in MND practice (Crockford et al., 2017). In our survey, it was determined that HCPs hoped to prevent causing distress to pwMND that they believed may come from identifying cognitive and behavioural changes. This barrier appears to be related to the task of breaking bad news. Bad news can be defined as delivering information that is going to drastically change the patient's view of their future (Buckman, 1984). Conveying information to patients that is sensitive or linked to a poorer prognosis is emotionally challenging for HCPs (O'Connell et al., 2024). At the same time, it is important that HCPs disclose information in practice as this aligns with the ethical principles of autonomy, beneficence and nonmaleficence (Edwin,

2008). Consequently, a HCP may be in the difficult position of knowing about cognitive and behavioural changes as being associated with MND but facing an ethical dilemma of delivering additional bad news to families, who may or may not prefer to learn about cognitive and behavioural changes. Further, studies have reported that HCPs experience increased difficulties when they are not able to provide solutions or hope (Carter et al., 1998). The difficulties HCPs describe in MND care are similarly reflected in literature which addresses the difficulties of conducting clinical conversations in life-limiting illnesses more broadly (Bernacki & Block, 2014). Specifically, if patients are having difficulty accepting their diagnosis, or experiencing distress related to the uncertainty of their prognosis (Bernacki & Block, 2014). As a solution to supporting HCPs manage these challenging conversations, incorporating evidenced-based methods of therapy such as motivational interviewing, solution-focused brief therapy and cognitive behaviour therapy (Marcus & Mott, 2014) may help to support clinicians to conduct these difficult conversations. To this end, the task of discussing cognitive and behavioural changes in MND may be seen as an opportunity for HCPs to provide education and check in with pwMND regarding their understanding about cognitive and behavioural changes in MND. Notably, respondents who held the view that pwMND would want to know about cognitive and behavioural changes, described that this knowledge would help pwMND to better understand the disease, feel prepared and in control. However, there is tension at play, as it is also suggested that HCPs may feel ill-prepared to conduct these difficult conversations (O'Connell et al., 2024 {Breen, 2020 #2443).

As previously mentioned, respondents reported no previous training in cognitive and behavioural changes in MND, and it is unclear whose professional role this task sits within. Further it is reported that few clinicians are trained specifically in communication techniques which may facilitate these difficult conversations and reduce patient distress. This again highlights an important opportunity to develop training and resources to support HCPs. Within the development of professional training modules, there is an opportunity to include

evidence-based communication training. Through developing HCPs skills, there is also a potential opportunity to change the perception of cognitive and behavioural changes in MND as being breaking bad news, and rather view these educational conversations as needed to support pwMND and carer's understanding of the disease and alleviate distress.

In contrast however, a facilitator to managing cognitive and behavioural changes in MND was that the majority of respondents held a belief in their capabilities. The TDF domain of *Belief about Capability* includes the constructs of 'perceived competence' and 'self-efficacy'. Within this study, respondents described feeling comfortable to address cognitive and behavioural changes with families. However, respondents also stated that they partly relied on carers to identify and report cognitive and behavioural changes, rather than instigate these clinical conversations as part of their usual practice. As such, there were variations on the frequency of how often clinical conversations about cognitive and behavioural changes took place, with some respondents suggesting that this was never conducted as part of routine clinical practice. Taken together, the reported inconsistencies in frequency and approach to discussing cognitive and behavioural changes highlight an additional complexity for identifying these changes. For instance, if carers are not routinely informed about cognitive and behavioural changes as being associated with MND by HCPS, it is possible that they would be unaware of what to look out for and hence not be able to report changes to the health care team. Considering the existing literature, which outlines that carers of pwMND experience high levels of carer burden (Chiò et al., 2005), and this burden has been shown to increase in the presence of cognitive and behavioural changes (Chiò et al., 2005), it appears even more important HCPs are equipped to ascertain what both the pwMND and the carer prefers for their care, how much information about the disease they individually want to know, and how the HCPs undertake clinical conversations to gain this information, so that they can then approach MND management with a tailored and individualised response. Therefore, through these tailored and individualised conversations, there is an opportunity to improve practice in relation to cognitive and behavioural changes, where

discussions about preferences and wishes are conducted in the early stages of the diagnosis. For instance, within palliative care it has been shown that earlier discussions about advancing disease facilitates patient choice in relation to their care trajectory, that is in alignment with their health care goals (Bernacki & Block, 2014). Overall, these conversations will increase a HCPs awareness of how much each pwMND, and carer wishes to learn about the disease. To this end, these supported conversations may in turn help families to feel supported to seek out information from their health care team. Furthermore, providing education on the types of behavioural changes that are associated with MND, may also prevent relationship breakdowns within the family, which may be caused by misunderstandings between carers and pwMND. Open communication about cognitive and behavioural changes may in turn also alleviate some of the burden felt by carers that has been described (Bernacki & Block, 2014; Visser et al., 2020). However, to undertake these conversations, it is important to consider the communication approach, as the communication needs of pwMND, who also have cognitive and behavioural changes, differ from those who do not.

Within the MND clinical guidelines, it is recommended that communication approaches be tailored to the cognitive capacity of each pwMND (National Institute for Health and Care Excellence, 2016). Notably, deficits in a pwMND's communication and cognitive capacity can create a barrier to participating in clinical conversations about their care. For instance, pwMND who also have cognitive and behavioural changes have been shown to have impaired decision-making capacity, difficulty processing new and complex information. In addition, a pwMND may have co-occurring language deficits (Paynter et al., 2019), such as word finding difficulties. These deficits may prohibit a pwMND from communicating effectively. As such, some pwMND may require more time to process and understand information (Fisher et al., 2017). Consequently, communication adaptations are necessary and need to be individualised and nuanced, based on an understanding of a pwMND's cognitive capacity. Consequently, HCPs need to first, be trained on how to adapt

communication to maximise a pwMND's ability to participate in clinical conversations, and then have access to evidence-based clinical care pathways to guide them to manage cognitive & behavioural changes in MND.

Clinical care pathways exist to support the implementation of evidence-based practice, provide support with planning and coordination of care, and define optimal sequencing to improve practices (Coffey et al., 1992). Additionally, clinical care pathway tools provide an opportunity to educate and upskill HCPs, as well as define responsibility and discipline-specific roles. Care pathways are important to consider, as HCPs have expressed in this survey that care planning in MND is often changing rapidly (McConigley et al., 2014) and is highly individualised due to the heterogeneity of disease presentations. In line with this, respondents highlighted the importance of relationship building and person-centred approaches as part of the care process to guide families through health care and management options to determine care preferences early before communication and cognitive changes may limit or prohibit this. It also became apparent that currently, providing information to families about cognitive and behavioural changes is hindered by unclear pathways and limited evidence of management strategies to back up these conversations. In addition to establishing optimal care pathways for pwMND, it is also important to involve families living with MND in developing care pathways for them in order to make these meaningful and relevant to the families that they are designed to help. Further, including families with MND who have also experienced cognitive and behavioural changes is vitally important to develop a better understanding of their specific care needs and to guide the development of these future clinical care pathways.

Limitations

While the survey results represent respondents from many disciplines, it is acknowledged that the findings are limited given this analysis is descriptive only. While respondents were from a broad range of disciplines, there were several disciplines that were represented by

only one or two respondents. This contrasted with a comparative over-representation of speech pathology respondents (n=19/55, 35%), creating skewed data that is likely to have affected the interpretation of the results. Consequently, these findings are not able to be applied across all disciplines involved in the care of pwMND and are not representative of the clinical approaches to cognitive and behavioural changes which may occur across all disciplines. More is needed to understand how cognitive and behavioural changes are approached within MDT care. This is especially important as neurology was most identified by respondents as responsible for addressing and managing cognitive and behavioural changes in MND, however there were only three respondents from neurology, limiting the interpretation and generalisability of these findings.

Further, respondents did not complete each survey question, resulting in missing data for some questions that may impact the interpretation. This also suggests that the survey length may have been too long.

A further limitation of the research is that the findings are skewed towards views of HCPs operating within the Australian health care context and hence may not be directly applicable to health care systems outside of Australia.

Future directions

This study documents that practice in relation to cognitive and behavioural change education and management in MND is inconsistent. Further, HCPs have not received training about cognitive and behavioural changes and additionally are not unclear if families wish to learn about this symptom of the MND. Hence, it is important to understand the views of people with lived experience of MND. Conversations with pwMND and their carers are needed to determine each person's preference for receiving information about, and the subsequent management of, cognitive and behavioural changes in MND. In addition, to develop a deeper understanding of the current state of clinical practice in relation to cognitive and

behavioural change in MND, interviewing HCPs may help to further unpack attitudes, and facilitators and barriers to practice. Such insights would help inform potential future approaches to reviewing and improving clinical pathways and in turn, support the production of training modules and resources for cognitive and behavioural changes to support HCPs.

Conclusion

This study showed that HCPs working with families experiencing MND have knowledge that cognitive and behavioural changes are associated with the disease and hold the view that these changes are important to manage. Additionally, HCPs felt somewhat comfortable conducting clinical discussions about cognitive and behavioural changes. However, barriers were identified related to training, resource availability, unclear scope of practice and a mixed belief if families with MND want to know about cognitive and behavioural changes. Clinical conversations about cognitive and behavioural changes are not occurring with consistency, and irregular screening practices were identified. Taken together, this may mean that identification and subsequent management of cognitive and behavioural changes in MND may be overlooked.

The previous chapter described the results of a mixed methods survey conducted with HCPs (ranging from many disciplines) who support families experiencing MND. To summarise this chapter, HCPs know cognitive and behavioural changes are associated with MND and most identified that managing cognitive and behavioural changes was important. Further, they described a level of comfort discussing this with families. However, there were inconsistencies in screening and management practices identified. Additionally, some HCPs held the belief that pwMND and their families would not want to know about cognitive and behavioural changes, which was viewed as a barrier to undertaking this clinical conversations. Therefore, to better understand factors which influence the attitudes, identification and management of cognitive and behavioural changes in MND, survey respondents were invited to participate in a follow-up semi-structured interview to elaborate on their answers. The interview questions derived for this study were centred around describing clinical context and MDT structures, the clinical pathways used to identify cognitive and behavioural changes, and subsequent management strategies that are implemented.

I interviewed twelve participants from a variety of disciplines and locations, both within Australia and overseas. The overarching aim of this study was to understand barriers and facilitators to managing cognitive and behavioural changes in MND. To aid sense-making and analysis, I used the TDF as a theory-driven framework to which I first deductively mapped the interview data, and then conducted further inductive analysis to identify patterns and categories within these data. The study is described in manuscript format below, with the intention of submitting to Translational Neurodegeneration.

CHAPTER 7: UNDERSTANDING PRACTICE: FACTORS INFLUENCING MANAGEMENT OF COGNITIVE AND BEHAVIOURAL CHANGES IN MND. A QUALITATIVE STUDY USING THE THEORETICAL DOMAINS FRAMEWORK.

Introduction

Motor Neurone Disease (MND) is an umbrella term for a subset of progressive neurogenerative diseases, including the most-diagnosed amyotrophic lateral sclerosis (ALS), progressive bulbar palsy (PBP) and rarer primary lateral sclerosis (PLS), and progressive muscular atrophy (PMA) (MND Australia, 2024b). Whilst the disease is well understood to impact on motor function and mobility, it is now also recognised that MND is associated with cognitive and behavioural changes (Pender et al., 2020). Up to 15% of people diagnosed with MND will also present with frontotemporal dementia (FTD) (Pender et al., 2020). Cognitive and behavioural changes in MND occur on an MND FTD continuum (Motor Neurone Disease Association, 2022) and up to 50% of pwMND experience changes (Pender et al., 2020). The most common cognitive changes are associated with executive functioning, language function and social cognition. Also, behavioural changes may present as apathy and disinhibition (Hill, 2016). Cognitive and behavioural changes can occur at any stage of MND progression, and in some cases have been found to occur prior to the development of motor function deficits (Rusina et al., 2021).

Whilst clinical guidelines for MND exist, they are lacking specificity and additionally have been reported to be of poor quality (Francis et al., 2023; Ou et al., 2023). Consequently, these guidelines may be difficult for HCPs to use to support their clinical decision-making. Information provided in these guidelines regarding cognitive and behavioural changes is general in nature, and provides a summary of their existence, rather than a toolkit with steps

for assessment and management. The MND clinical guidelines from The National Institute for Health and Care Excellence (NICE), published in 2016, report that HCPs should be aware that cognitive and behavioural changes are associated with MND, and this should be factored into clinical management. However, these guidelines stop short of providing specific assessment and management strategies, but rather recommend seeking further information from the mental health capacity act and dementia literature. Additionally, within these documents, the referenced sources of additional information are not derived from evidence specific to MND, and often sit within the dementia literature. Taken together, these management approaches to cognitive and behavioural changes in MND may be of limited use in MND, as dementia decline is often associated with decline in memory and overall cognitive skills, which progress to levels of severity which impact on a person's ability to be autonomous in activities of daily living (Emmady et al., 2020). Cognitive changes in MND, however, overlap with the FTD syndrome (Bak, 2010) and are less associated with memory deficits. Rather, memory changes that may be described in both FTD and MND have been shown to be associated with executive dysfunction and language decoding, rather than impaired recall (Abrahams et al., 2005).

Detection and the subsequent management of cognitive and behavioural change in MND is important, as these changes are associated with reduced treatment uptake, reduced treatment adherence, reduced quality of life for pwMND (Radakovic et al., 2024) and increased caregiver burden (Oseland et al., 2023). However, it remains unclear if HCPs who work with pwMND are identifying, and subsequently managing, cognitive and behavioural changes in a consistent way, or even at all. Where these practices are described, many inconsistencies have been reported (Crockford et al., 2017) (Chapter 6). For example, whilst HCPs have described observing cognitive and behavioural changes and acknowledged that management as important, regular screening is rarely conducted as part of usual clinical practice (Chapter 6). Further, inconsistencies in management are also reported (Chapter 6). Adding to this complexity, there is limited information available to guide HCPs to manage

cognitive and behavioural changes that is derived from MND-specific evidence (Francis et al., 2023).

Previous investigations have identified several implementation barriers for cognitive and behavioural change screening. These barriers are related to a lack of resources, staff attitudes to screening, and the HCP's beliefs that families experiencing MND do not want to know about cognitive changes (Crockford et al., 2017). These barriers have been shown to inhibit practices related cognitive and behavioural change (Crockford et al., 2017).

An approach to support better understanding of the barriers that influence clinical practices, is to incorporate theories of human behaviour and behaviour change within research. Using a theoretical approach helps to conceptualise complex issues, gives greater depth of interpretation, adds consistencies and comparability, increases relevance and aids in knowledge translation (Atkins et al., 2017; Cane et al., 2012). One such framework is the TDF, which was initially developed to identify factors which influence health professionals' knowledge, skills and behaviours (Michie et al., 2005).

Underpinned by theoretical understanding of behaviour and behaviour change, the TDF consists of fourteen validated domains (Cane et al., 2012). To support the implementation of new (evidenced-based) practice recommendations and/or to change existing practices, the TDF allows for the systematic assessment of barriers and facilitators at the individual, team and organisational level. The TDF can be used prospectively to assist with research design or as was the case for this research project, as an interpretive framework to better understand behaviour and to identify potential barriers and enablers to implementing management of cognitive and behavioural change in MND. This framework will be applied to answer the following research question:

What factors influence management of cognitive and behavioural changes in MND?

Method

Research aim

The aim of this study was to explore HCPs' experiences of addressing and managing cognitive and behavioural changes in MND, including individual factors which influence clinical approaches to identifying and managing this symptom. Further, we explored potential facilitators and barriers to management of cognitive and behavioural changes in MND, as well as HCPs' behaviours and attitudes related to their clinical practice.

Study design

As the study was exploratory, we employed a qualitative design consisting of semi-structured interviews. This method was chosen as it allows for a deep exploration of each participant's thoughts and experiences, and enabled us to employ a flexible interview protocol, which consisted of follow-up questions, probes and comments. Through this, we were able to explore each participant's thoughts, feelings, experiences and beliefs, and from these, which influence approaches to, addressing and managing cognitive and behavioural changes in MND. Subsequently, the interview guide (Appendix 8) consisted of open-ended questions, designed to elicit information about each participant's individual clinical experiences and beliefs.

The interview guide for this study was created after analysis of the survey data was complete (Chapter 6). This approach allowed the research team to identify areas within the survey findings which required further probing and exploration in relation to the identification and clinical management of cognitive and behavioural changes in MND. The interview questions were developed to enable participants to provide more information about current clinical contexts, management approaches and personal factors which may be at play. Table 20 shows the question development process. The interview questions and topic guide were reviewed and refined iteratively within the research team. The interview questions explored the practice context, and clinical approaches to managing cognitive and behavioural

changes in MND. Specifically, participants were asked to describe previous training, screening practices, management strategies, beliefs and attitudes related to cognitive and behavioural changes in MND. Once finalised, an amendment to the existing ethics approval was submitted and the interview topic guide was provided (Proposal #4660) (Appendix 01 and 8).

Table 20.

Health care professional interviews (Chapter 7). Question development process

Survey data	Research question	Semi-structured interview – guiding question
70 % of HCPs conduct a screen for cognitive and behavioural changes at least some of the time.	1. What do you do to manage cog and behavioural in your routine practice? 2. What are the care pathways for managing cog and behavioural change in MND?	1. Can you describe what might happens after this screen has been conducted in your clinical setting?
24% of HCPs felt somewhat uncomfortable and 33% felt somewhat comfortable talking about cognition with families.	1. How do HCPs feel about discussing cognitive and behavioural changes with families experiencing MND?	1. From your perspective why would HCPs feel less or more comfortable talking about cog in MND? 2. What do you think would improve this for HCPs? Prompt – in your experience what makes discussing this more uncomfortable? More comfortable? Prompt – in your experience how do they receive this information? How do you find they generally react to learning about this symptom? If they have been screened to have change, how do you find they react to the news they have experienced cognitive/behavioural change? 3. Do you think stigma exists in this space?
45% of respondents provided education for cognitive and behavioural change associated with MND, and much of this education (over 34%) was provided verbally only.	1. How do HCPs educate pwMND about cognitive and behavioural change?	1. Can you tell me about the provision of education related to cognitive and behavioural change in MND specific to your setting? Prompt - In what medium is this sharing of information done?

Survey data	Research question	Semi-structured interview – guiding question
respondents in the survey described a multi-disciplinary approach to managing cognitive/behavioural deficits in MND.	1. What is the structure of a multidisciplinary team in MND?	1. What does MDT mean/look like in your setting? Prompt – Disciplines? /Barriers? /Processes?
Approx. half the respondents in the survey report having received training for supporting someone with cognitive/behavioural changes related to any neurodegenerative disease.	1. What training have HCPs received about cognitive and behavioural changes in MND? 2. What training do HCPs want to undergo to support families living with MND who also are experiencing cognitive and behavioural changes?	1. Have you received training about cognitive and behavioural changes in MND? If Yes - Can you describe this? If No – Can you suggest why this might be? 2. If you were to design a training program for cognitive and behavioural changes n MND, what do you think needs to be included?

Participants

Participants were HCPs from any discipline and any location, who provided care for families experiencing MND. All participants were recruited via a previous survey (Chapter 6) and as such, all participants in this study indicated that cognitive and behavioural changes are associated with MND in their previous survey responses.[9]

Specifically, respondents who completed the survey (Chapter 6) were invited to voluntarily participate in a follow-up interview. Any HCP who worked with an MND caseload, from any location, discipline or practice setting was eligible to participate. Each respondent from the survey who provided their contact information was individually emailed to ask if they agreed to participate in an interview and sent a participant information and consent form (PICF) as per Appendix 10. Once participants provided informed consent, an interview time was scheduled.

Semi-structured interviews

A single online video interview was conducted at a time convenient for each participant. Interviews were conducted in Microsoft Teams and recorded with participant's consent, to enable an audio file for transcription purposes to be created. All participating HCPs were provided with a gift card to the equivalent value of AUD $40.

Data analysis

For each interview, the HCP information was de-identified, and each participant was assigned a project code to aid in data analysis. All interview data were then transcribed verbatim using an external transcription service. The completed transcriptions were error

[9] Note: The inclusion criteria of the survey included a question which asked respondents if cognitive and behavioural changes were associated with MND. If a respondent answered "No" to this question, they were excluded from participation in the survey, and subsequently also from participating in the study described in this chapter.

checked against the recording for accuracy by the candidate and then imported into NVivo software for data analysis.

Initially, all interview transcripts were reviewed, undertaking a process of data immersion and data familiarisation. During this stage, memos were recorded to both summarise thinking and to capture the setting and candidate's feelings as they related to the research questions.

The initial coding process was conducted deductively, where data were coded to the revised version of the TDF (Atkins et al., 2017; Cane et al., 2012; McGowan et al., 2020; McSherry et al., 2012). Table 21 describes the TDF domains, descriptors and constructs that contribute to each domain. To organise the data, a constant comparative method of analysis was undertaken to revise and refine the data within the TDF domains (Glaser & Strauss, 2017; Glaser et al., 1968). To support this process annotations and memos were created to support thinking and for later sense checking amongst the research team.

Table 21.

The theoretical domains framework domains with corresponding definition and constructs. Taken from Cane et al. (2012)

Domain and definition	Constructs
1. Knowledge (An awareness of the existence of something)	Knowledge (including knowledge of condition/scientific rationale) Procedural knowledge Knowledge of task environment
2. Skills (An ability or proficiency acquired through practice)	Skills Skills development Competence Ability Interpersonal skills Practice Skill assessment

Domain and definition	Constructs
3. Social/professional role and identity (A coherent set of behaviours and displayed personal qualities of an individual in a social or work setting)	Professional identity Professional role Social identity Identity Professional boundaries Professional confidence Group identity Leadership Organisational commitment
4. Beliefs about capabilities (Acceptance of the truth, reality or validity about an ability, talent or facility that a person can put to constructive use)	Self-confidence Perceived competence Self-efficacy Perceived behavioural control Beliefs Self-esteem Empowerment Professional confidence
5. Optimism (The confidence that things will happen for the best or that desired goals will be attained)	Optimism Pessimism Unrealistic optimism Identity

Domain and definition	Constructs
6. Beliefs about Consequences (Acceptance of the truth, reality, or validity about outcomes of a behaviour in a given situation)	Beliefs Outcome expectancies Characteristics of outcome expectancies Anticipated regret Consequents
7. Reinforcement (Increasing the probability of a response by arranging a dependent relationship, or contingency, between the response and a given stimulus)	Rewards (proximal/distal, valued/not valued, probable/improbable) Incentives Punishment Consequents Reinforcement Contingencies Sanctions
8. Intentions (A conscious decision to perform a behaviour or a resolve to act in a certain way)	Stability of intentions Stages of change model Transtheoretical model and stages of change

Domain and definition	Constructs
9. Goals (Mental representations of outcomes or end states that an individual wants to achieve)	Goals (distal/proximal) Goal priority Goal/target setting Goals (autonomous/controlled) Action planning Implementation intention
10. Memory, attention and decision processes (The ability to retain information, focus selectively on aspects of the environment and choose between two or more alternatives)	Memory Attention Attention control Decision making Cognitive overload/tiredness
11. Environmental context and resources (Any circumstance of a person's situation or environment that discourages or encourages the development of skills and abilities, independence, social competence and adaptive behaviour)	Environmental stressors Resources/material resources Organisational culture/climate Salient events/critical incidents Person × environment interaction Barriers and facilitators

Domain and definition	Constructs
12. Social influences (Those interpersonal processes that can cause individuals to change their thoughts, feelings, or behaviours)	Social pressure Social norms Group conformity Social comparisons Group norms Social support Power Intergroup conflict Alienation Group identity Modelling
13. Emotion (A complex reaction pattern, involving experiential, behavioural, and physiological elements, by which the individual attempts to deal with a personally significant matter or event)	Fear Anxiety Affect Stress Depression Positive/negative affect Burn-out
14. Behavioural regulation (Anything aimed at managing or changing objectively observed or measured actions)	Self-monitoring Breaking habit Action planning

Once the initial deductive coding was complete, inductive analysis was undertaken to further fracture the data situated within each domain of the TDF into subthemes, and to identify patterns and categories (Figure 29).

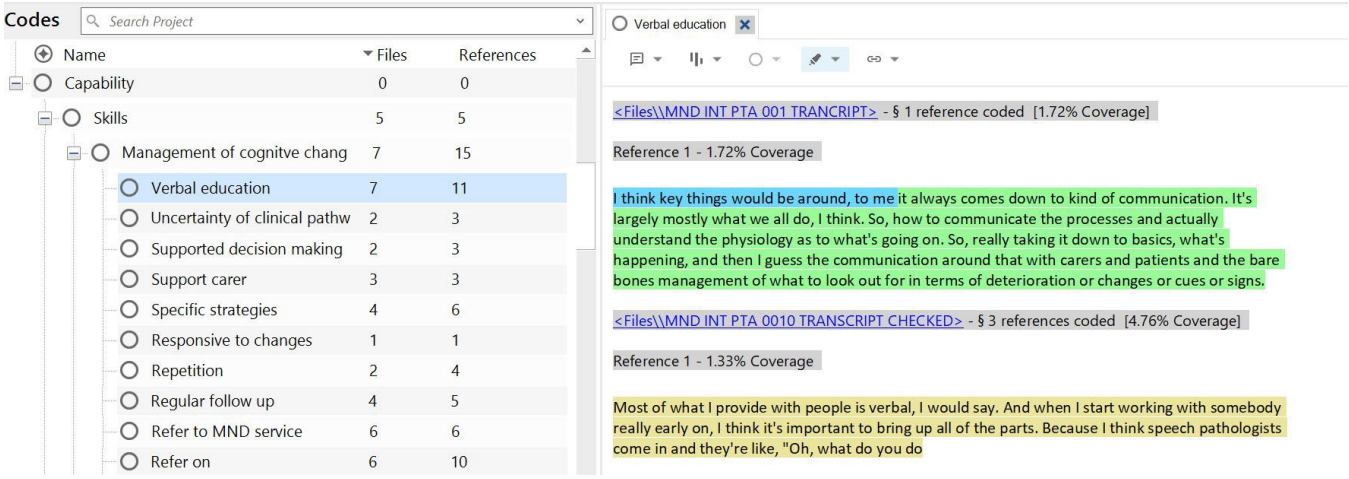


Figure 29: Data analysis in NVivo. Initial coded consisted of deductive analysis to the TDF Domains followed by inductive coding within each domain to identify patterns and categories. ***(TDF: Theoretical Domains Framework)***

Then, after the initial round of inductive coding within each TDF domain was complete, these newly generated subcodes were discussed with the broader research team and a further round of inductive coding was undertaken to identify broader patterns within the data and refine themes. During this process, key points for each subtheme were recorded as shown in Table 22.

Table 22.

Deductive Inductive coding of raw data to Theoretical Domains Framework domains

Excerpt of raw data	Example of researcher annotation	Deductive coding to TDF domains	Inductive coding	Theme
This may never happen to you, this is not about you, but this is what can happen	*respondent describes providing education that is depersonalised*	Skills	Management - Verbal education	Approaches to management of cognitive and behavioural changes in MND
But in terms of other written resources that's provided, what we normally give is what's available on the MND Australia website, and it's only basic, really basic information	*respondent descries using MND services to access information for clients*		Management - Written education	

Findings

Participants

Thirteen participants (three speech pathologists, two occupational therapists, one respiratory physiotherapist, two physiotherapists, one respiratory nurse, one clinical neuropsychologist, one MND support coordinator, one palliative care nurse and one MND support service manager) were interviewed as shown in Table 23. Of the 13 participants, four were located in Queensland, three in South Australia, and one each in Victoria, New South Wales, the Australian Capital Territory, New Zealand and the UK. The average length of each interview was approximately 40 minutes.

It is noted that one participant was a clinical neuropsychologist. Their clinical context included conducting neuropsychological assessments with people who have a diagnosis of a degenerative neurological condition, as well as providing training to other members of their MDT about cognition and behaviour. Therefore, given the specific nature of their university education and clinical work centering around cognitive and behaviour, their foundational level of knowledge differed from the other participants within this study. The interview data from this participant were included for analysis in the same way as all other participants.

Table 23.

Characteristics of participants.

Participant	Discipline	Service model	Numbers of years working in MND	Number of pwMND supported	Location
PTA001	Respiratory physiotherapist	Community	1-5	20-99	SA
PTA002	Clinical neuropsychologist	Outpatient clinic	5-10	20-99	VIC
PTA003	Support Coordinator	Disability case management	5-10	100+	SA
PTA004	Physiotherapist	Community	5-10	100+	QLD
PTA005	Physiotherapist//MND Clinic Coordinator	Hospice	5-10	20-99	UK
PTA006	Occupational therapist	Community	10-15	20-99	NSW
PTA007	Occupational therapist	Outpatient clinic	1-5	5-20	ACT
PTA008	Speech pathologist	Outpatient clinic	1-5	20-99	QLD
PTA009	Speech pathologist	Community	1-5	5-20	SA
PTA010	Speech pathologist	Community	5-10	20-99	SA
PTA011	Respiratory nurse	Community	10-15	100+	QLD
PTA012	Palliative care nurse	Community	5-10	100+	QLD
PTA013	MND Support Service Manager	Community	20+	100+	New Zealand

Note: (PTA: participant identification code; MND: Motor Neurone Disease, SA: South Australia; VIC: Victoria; QLD: Queensland; UK: United Kingdom; NSW: New South Wales; ACT: Australian Capital Territory)

The following six overarching themes were derived:

i. *Knowledge of cognitive and behavioural changes in MND*,

ii. Clinical context is both a facilitator and barrier to managing cognitive and behavioural changes in MND

iii. Approaches to managing cognitive and behavioural changes in MND

iv. Overwhelming families living with MND

v. Reflecting on changing practice to manage cognitive and behavioural change in MND

vi. Creating professional development for cognitive and behavioural changes in MND

Within each theme, relevant TDF domains are described to assist in the understanding of the barriers and facilitators to managing cognitive and behavioural changes in MND in clinical practice. Quotes are also used to illustrate these findings and provide additional context within each theme.

i. Knowledge of cognitive and behavioural changes in MND

This theme describes participants' knowledge about cognitive and behavioural changes associated with MND and includes specific descriptions of their clinical experiences. Within this theme, the TDF domains of *Knowledge, Social influences, Skills* and *Belief about capabilities* are captured.

Whilst all participants had knowledge that cognitive and behavioural changes may be associated with MND, some believed that the general community viewpoint was that cognition and behaviour is spared in MND. Participants suggested that this may be because knowledge about cognitive and behavioural changes associated with MND is a relatively recent field of enquiry, given the disease was previously only thought to impact on motor function (hence the name Motor Neurone Disease).

> But I do not think a lot of people quite understand the condition itself, or that there's sort of cognitive and behavioural decline that comes with it from a general population perspective… and to be fair to myself as well, I was not aware until I started working in the space about the cognitive and behavioural factors that come with it (Participant 01).

> When I first started working with MND, it was still thought to not affect cognition at all (Participant 06).

> I would guess that it again, it's a relatively new area, which some people have had a lot more education on than others, perhaps. And if people do not feel they understand it themselves particularly well, then it's going to be less easy for them to bring it up (Participant 05).

Some participants suggested that they themselves sometimes assume that pwMND do not experience cognitive and behavioural changes, and this may not be considered until there are overt signs.

> …I guess you immediately assume that an MND patient will be cognitively intact. So, you speak to them like they are, and you give them information like they are. And you never really consider that they might have cognitive and behavioural changes until you can physically see it yourself (Participant 08).

> I think with MND, there is that sort of quintessential thought. It's the mind does not get impacted by MND, it's just the body. And I think maybe with that being sort of a narrative over the years of the past, maybe there is that thought from the community that it's a bit extra if you've got this extra in cognitive impairment (Participant 06).

Participants also provided a description of cognitive and behavioural changes, with lack of insight, impaired decision-making, rigid thinking, and impaired empathy described. Only one participant described apathy (sometimes termed initiation apathy) as a clinical presentation of cognitive and behavioural changes associated with MND.

> It's just that lack of insight in decision making process, and not considering the big picture stuff, particularly not caring about their carers. They want it done this way, so they're going to do it this way, regardless. And not looking at the impact that has on everybody else (Participant 11).

> So, for our palliative care patients with MND, most of the time the things we're looking at are the frontotemporal dementia or the rigiditive thinking (Participant 12).

> I can only think of one patient I've had in my entire career who's been so impaired that she was not able to make any decisions for herself at all. Sometimes people just need a bit of support and a bit of chatting with the family about. Do you feel that this is a decision they would've made before their diagnosis, and before their personality changed, and their thinking changed? And normally the answer is, Actually, yes. They've always been quite determined, and stubborn, and fixed in their thinking and it's just more of an exaggeration of their normal personality, quite often, I find (Participant 05).

Additionally, whilst participants reported knowing that cognitive and behavioural changes are associated with MND, they described learning about cognitive and behavioural changes from within the broader dementia literature.

> I mean, I have received training in regard to dementia management back when I was a new grad or very early on in my career, but I have not received any specifics (Participant 01).

> Oh, yes, I reckon I've done some training over the years around Alzheimer's and dementia, cognitive communication changes. Actually, not neurodegenerative, but relating to stroke. No, not specifically. Oh, sorry, I've done a bit, also not related, but around clients with TBI and those cognitive communication difficulties (Participant 10).

One participant described undertaking a training course provided by an MND Association. This consisted of a self-directed online training module about MND more broadly, which included information on cognition.

> I have received training. I do not know if you could call it formal training though. We had a training course that was developed within The MND Association…So they had this 27-volume program that you could work your way through. It was predominantly online, and there was a large portion of that was cognitive as well (Participant 03).

All participants shared the view that each pwMND experiences the disease differently due to the heterogenous nature of MND and the variations in functional deficits. Given the continuum of cognitive and behavioural changes that can occur in MND, there are variances both within and across individuals, and one participant provided valuable insight into these complexities:

> I wonder if people struggle with the nuances sometimes…and I think people do not quite know what it looks like because they do not maybe have that nuanced

> understanding of cognition…people do not know what to say because they sort of think, I do not know what your cognition's going to look like, because they've seen every variation along the spectrum. So, I think that the difficulty in predicting what might happen to people is quite hard (Participant 02).

As such, participants described gaps in their knowledge about cognitive and behavioural changes in MND with uncertainty related to prognosis, pathophysiology, and uncertainty related to management of cognitive and behavioural changes was highlighted. Given this, the ways that participants have constructed their knowledge of cognitive and behavioural changes in MND is related to the ways this has presented in the people they have worked with. Consequently, participants recognised that knowledge gaps about how cognitive and behavioural changes can manifest within and across pwMND, and an uncertainty of when is the best time to discuss this with families undermines their capability to feel confident when speaking about this with families.

> Think a lot of HCPs feel uncomfortable talking about something that they do not have an adequate knowledge of. I know that I do not often have any issue talking about a difficult topic with a patient if I have the knowledge and the solutions to back myself up, if that makes sense? So, I think I probably put that there would be some level of discomfort for myself bringing a topic up like this because I do not feel my knowledge is adequate to back myself up if the family member or the patient had specific questions (Participant 08).

> In terms of cognition and the changing of behaviours. From my perspective, I find it's quite a difficult conversation to have because I do not know. I think maybe more that knowledge gap. Knowledge gap or everyone is so different, and I feel like a bit of a politician almost when you answer some questions about, I cannot

> really say. I can give a general response back to what you're asking. if that sort of makes sense (Participant 01).
>
> Yeah, I guess one, what's the progression? Two, what could that look like, how do you manage that, what are the timeframes? I think the timeframe thing is really what tends to come up, one, about the sort of prognosis and deterioration in general, but also in terms of cognition and the changing of behaviours. From my perspective, I find it's quite a difficult conversation to have because I do not know. I think maybe more that knowledge gap. Knowledge gap or everyone is so different, and I feel like a bit of a politician almost when you answer some questions about, I cannot really say. I can give a general response back to what you're asking. if that sort of makes sense. I cannot be direct in saying, this is why it's occurring or why this time, what the timeframe will look like, or anything like that (Participant 01).

In summary, participants know that cognitive and behavioural changes are associated with MND, however, they described gaps in their knowledge which impacts on their confidence to discuss this with families. Further, participants believed there was a lingering perception in the general population that cognition is spared in MND, and consequently these changes may not always be considered. Participants also described that because of this perception, HCPs may assume that a pwMND has intact cognition, unless alerted otherwise.

ii. Clinical context is both a facilitator and barrier to managing cognitive and behavioural changes in MND

The theme of Clinical context relates to the clinical settings and scope of practice that participants described being employed within to support families with MND. Here, the TDF domains relating to this theme are *Environmental context and resources, Skills,*

Social/professional role and identity, Belief about capabilities, Optimism and *Beliefs about consequences.*

When asked to describe their clinical context, all participants described a variety of MDTs and clinical models, with a range of disciplines from both medical and allied health being part of their usual team. These MDT services and disciplines are summarised in Table 24.

Table 24.

Multidisciplinary team (MDT) compositions as described by participants.

Service	Frequency	Disciplines
MND outpatient clinic	Monthly	Physiotherapist, (clinic coordinator), palliative care, occupational therapist, speech pathologist, dietician, clinical nurse, doctor, respiratory physiologist, psychologist, counsellor orthotics.
Community outreach chronic disease team	Not described	Neurologist, respiratory specialist, nurse, occupational therapy, dietician, physiotherapist.
Community – in home service or outpatient clinic	Not described	Respiratory physio. No direct MDT described.
Support coordination	Not described	Occupational therapy, support coordinator (nurse)
Progressive neurological diseases inpatient unit integrated with outpatient clinic		Physiotherapist, palliative care, occupational therapist, speech pathologist, dietician, social work, clinical psychologist, neuropsychologist, neurologists, psychiatrist, nurse, pastoral care, music therapy, diversional therapy.
Chronic disease service – community	Four to eight weeks	Over 65s Respiratory nurse, physiotherapist, occupational therapy, dietician, social work Under 65s Dietician, social work psychology.
Community Palliative Care	Not described	Palliative care nurse
Community Health	Four to six weeks	Palliative care occupational therapy, physiotherapist, specialist palliative care doctor.

Service	Frequency	Disciplines
Outpatient rehabilitation clinic	Monthly	Occupational therapy, physiotherapy, speech pathology, dietician, social work MND advisor
MND clinic	Not described	Neurologist, speech pathologist, neuropsychology
Private practice	Not described	Speech pathologist
Private practice	Not described	Support coordinator, speech pathologist, dietician

Clinical context facilitators

Participants identified that working within MDT is a facilitator to managing cognitive and behavioural changes in MND. Further, they expressed optimism in this team-based collaborative approach to care for families with MND and felt confident and clear about the roles and responsibilities of others within their MDT. Participants believed they were supported by their colleagues and described relying on them for information about cognitive and behavioural changes. A frequently described method for discussing MND patient care was through regular MDT case conferences, with collaborative approaches to care described. Participants believed that these case discussions provided them with an avenue to discuss their concerns about cognitive and behavioural changes, amongst the greater team. Here, they described skills in developing a consensus, whether changes had been observed by others or not, and ways they approach management.

> We have a MDT meeting which is run by me and the physio and the respiratory CNC at the hospital. And that's sort of a forum where we chat about patients…and their behaviour as a part of that, if any changes in their cognitive or behavioural sort of functioning…and I guess that's a forum where everybody who does attend, which is pretty much everyone that could be involved, can talk about any issues with that. And I guess in terms of the management it is, what's the impact of that and what's the distress level, have we linked in? (Participant 06).

> …we tend to discuss our thoughts and feelings, and we do not often screen to be honest. It’s quite unusual that we do screen. And we tend to just go with how the patient is presenting, and we just manage them depending on the symptoms they are showing (Participant 05).

> We case conference every month, to see who's going to the clinic, and then talk about any other clients at the same time (Participant 11).

Clinical context barriers

It was less clear who, within the MDT, is responsible for providing education about and management for cognitive and behavioural changes to families. Specifically, participants described a level of discomfort when a pwMND is referred to them for follow up for cognitive and behavioural changes, believing this is not part of the scope of their discipline.

> I know a lot of people kind of refer to me and I'm like, eh, I cannot really help with that (Participant 01).

Others perceived that cognitive and behavioural change management sits within the scope of occupational therapists or neurologists.

> The OTs would monitor. They would be the discipline that is designated to monitor the deterioration as time goes on and it will be fed back to the multidisciplinary team who's involved in looking after that patient in terms of how we go about supporting the family the best that we can. So that's as far as what we would do with the assessment findings that we gathered (Participant 05).

Additionally, participants expressed that a primary barrier to managing cognitive and behavioural changes in MND related to *Environmental context and resources*. Most participants described working at capacity and having little additional time to participate in professional development. Further, limitations to clinical appointment time, insufficient funding, geographical distance from patients, and an inability to work to their full scope of

practice mean that other symptoms of MND are prioritised for care, over cognitive and behavioural changes.

> This service is also a service that does not have designated or dedicated funding. It comes out of the ** pool.[10] And the reason for that is because we have a very passionate ** who is a strong advocate for the MND consumer (Participant 04).

> I think there's, part of it would be, okay, where are we up to with the latest literature, like guidelines, and having some allocated space to look at those? Because I think we might get five, 10 minutes to have a quick click and look at stuff, but you do not get time to spend a whole day looking at stuff, and researching stuff, particularly (Participant 11).

> …Nothing happens next, and this is a real frustration. We're a small place with a very low number of pwMND. We cannot refer on to the memory clinic until we have a proper diagnosis…but the problem is there's a waiting list of about six months to a year. So, they always die before they ever get an appointment. So, they never get a neuropsychology assessment (Participant 05).

> I guess the challenge is that I get eight hours a week…so sort of giving that support to the carers within that timeframe and the geographical challenges to it as well makes it difficult (Participant 12).

In summary, this theme described both facilitators and barriers related to the clinical contexts that participants are working within to manage cognitive and behavioural changes in MND. Here, participants were confident in their capability and skills to manage symptoms of MND

[10] ** denotes the removal of information to protect anonymity

that are situated in their scope of practice. Whilst varied multidisciplinary clinical models were described, most felt confident with the support they received from their peers. However, participants described barriers to managing cognitive and behavioural changes related to both a lack of resources and an unclear clinical ownership for managing cognitive and behavioural changes. Participants believed the lack of resources and a need to prioritise other clinical and practice demands impacted negatively on both their ability to support their caseload and to develop their professional skills and knowledge.

iii. Approaches to managing cognitive and behavioural changes in MND

This theme relates to cognitive and behavioural management strategies that participants described enacting. Management in this context encompasses education, assessment or screening and specific strategies to support both a pwMND and their carer. The TDF domains that relate to this theme are *Belief about capabilities, Skills*, and *Belief about consequences*.

Providing education to families was a large component of what participants do clinically to manage cognitive and behavioural changes in MND. Education was provided both verbally and in writing. Some participants also believed that education should be presented in a depersonalised way, early in disease progression.

> It is verbal because I think a lot of the education, what's labelled as education is we just highlight the changes that the person will be going through like difficulties with concentration, maybe lacking in executive thinking and functioning, and sometimes highlighting that it is very easy that through learned behaviour and through patterns that they can mask these changes. So that would be classified as education that the family would be getting, and the patients would be advised. But in terms of other written resources that's provided, what we normally give is

> what's available on the MND Australia website, and it's only basic, really basic information. So, I sort of check in with people around how they feel about what information they want. But for a lot of people, I find that if you give them information that is depersonalised, this may never happen to you, this is not about you, but this is what can happen, it's much more easily received (Participant 01).

> Obviously, there's a fact sheet that we do provide from the MND Australia organisation I guess we just give out the handout and field any questions that may come of it, link them in with people that have a bit more time to sit and field ponderings from the patient and their family and the broader family. It's more about education (Participant 06).

Further, participants described educational conversations need to be responsive to individual differences and tailored to individual needs. Here, most participants described feeling confident in their own clinical ability and suggest that this skill is learned over time, which builds with experience.

> I would assume that most HCPs are comfortable telling people before anything has eventuated, this can happen with MND. I think that is fairly straightforward. And then, when you get people who are having these significant cognitive changes, it's really apparent to everybody that that's going on (Participant 10).

> Having done this for eight, nine years, it does not faze me anymore, because you see everything. You still get new scenarios that come, that you have not seen…. (Participant 11).

> I think any conversation with patients is a learned skill. I do not think it's something that we have, and I think it's experience and learning from senior

> members of team and so forth. And so, I think having those conversations and the way you approach it is probably if you approach it in a manner, you might just get shut down and so the conversation does not continue on. So, I guess it's just a means of how we approach that conversation that is not offensive. If we're talking to a patient about increasing confusion or dementia and forgetfulness, if they're not aware of that or the family's picking up on it more, not so much to the patient. I guess it's a matter of putting it in a phrase that you're still quite respectful to the patient to figure out whether they're acknowledging these things or not (Participant 12).

When reflecting on the literature that is referenced to guide practice related to cognitive and behavioural changes in MND, participants described drawing on strategies derived from other neurodegenerative diseases, such as dementia. Participants also acknowledged that there are additional complexities of managing cognitive and behavioural changes in MND due to the often-severe functional deficits in both mobility and communication. Here, they expressed their belief that clinical conversations need to be personalised and nuanced to each person/family.

> Well yeah it is coming from the dementia space, I'd say more generally. But when it comes to MND, the specifics tend to be around modifying things for the other issues the person has…how can we support them in the context of the cognitive changes they are also experiencing…So yes, it's not so different from some other progressive neurological diseases, but there is complexity that comes with the other changes in MND for sure (Participant 02).

Regular screening or assessment practices were rarely described as part of routine clinical management for cognitive and behavioural changes in MND. Instead, participants relied on family reports or a consensus approach to determine cognitive and behavioural changes.

> So, if we do suspect it, I guess it often comes from observation or from a family member who rings in and says this is happening, we give them information (Participant 06).

> I guess we are very reliant on family to provide this information to us as well. And I guess maybe that could be where a gap is, because if they're not aware that this is something that can happen for pwMND, well then, it's not going to be brought to our attention to be looked at either…As a team, we tend to discuss our thoughts and feelings, and we do not often screen, to be honest. It's quite unusual that we do screen. And we tend to just go with how the patient's presenting, and we just manage them depending on the symptoms that they're showing (Participant 12).

> We do not do any specific cognitive screening. It would probably rely on either myself or our neurologist, (name omitted) picking up on something anecdotally in clinic (Participant 08).

When reflecting on the processes used to identify cognitive and behavioural changes, one participant highlighted that in the presence of dysarthria or anarthria, it can be difficult to differentiate or determine via clinical observations if cognitive and behavioural changes are present:

> But I've had clients where it's almost so mild, and between the fact that it is really mild, and then between their communication difficulties, maybe they're using eye

> gaze already, and it's so subtle that it's hard to tease out whether it is really a cognitive impairment (Participant 10).

Participants described selecting management strategies for cognitive and behavioural changes that are unique to the needs and presentation of each individual. For example, adapting communication and using communication aids to support understanding:

> So, if they have difficulty understanding, then we do paraphrasing. We use images. We use communication boards, use the tablet, things like that (Participant 04).

> So, I guess it's very needs based and unique for each (Participant 12).

> And when you have a look at it, and you notice that they kind of do not fall in that normal category, it's not about treating everyone the same…It's really about figuring out what their main, trying to find the right word, idiosyncrasies. Just the changes of behavior, the main ones of note, and how we can manage it per person, per issue that develops (Participant 03).

Participants also explained several practices they undertook to adapt their clinical routine in the presence of cognitive and behavioural changes. These strategies included prior reading of case information and phone calls to families to conduct pre-appointment information gathering about the pwMND. Through these different strategies to gather relevant information, participants described making adaptations to individually support the pwMND in the best possible way. One participant described that their aim is to maximise a person's capacity to be involved in decision making about their own care. Further, they described that

cognitive and behavioural changes do not automatically negate a person's ability or capacity to be involved in care discussions.

> We'd always want to be working with a supported decision-making sense. You do not want to be taking away someone's capacity unnecessarily. So, the default is everyone has capacity until there's a reason to think otherwise. So, if someone has cognitive impairment but we think they can still participate in those decisions, let's say it's around medical decisions, or about lifestyle decisions, then we want to maximise their ability to participate (Participant 02).

In summary, participants described feeling confident in their clinical skills. Most participants described drawing on strategies for cognitive and behavioural changes from evidence derived from dementia research. Participants described individualising care and adapting practice in response to learning about cognitive and behavioural changes.

iv. Overwhelming families living with MND

This theme describes the barriers identified for managing cognitive and behavioural changes in MND that are related participant's concerns of overwhelming families and causing distress. The TDF domains of *Social influences* and *Belief about consequences* relate to this theme.

Most participants described not wanting to overwhelm families with MND, as they are already coming to terms with both a terminal diagnosis and the severe functional impacts of the MND. As such, participants believed that introducing information about cognitive and behavioural changes would be received by families as another burden that they would need to face. Additionally, due to the variability of the disease, participants believed that it may be

difficult to address cognitive and behavioural changes, as not all pwMND will experience these changes.

> Having the conversation saying, this could happen. And unfortunately, you are showing obvious signs of some of this stuff, so we need to look at it further. That's where the conversation gets harder because you... They're already aware they've got a terminal illness and now you're telling them, so not only physically you're going to be struggling. Now mentally things are going to be crap as well. And that's the hard conversation. It's just like kicking a guy when he is down (Participant 3).

> Because I genuinely do think, in this population, that cognitive changes are really important, but I think we need to also be careful of not adding to the load of these clients, emotionally and timewise, when they're already dealing with a lot (Participant 10).

> Not everyone with MND gets all of the same symptoms, and so I guess you do not want to overload people with too much information and scare them off about potentials that could happen. But at the same time, I guess it's good for them to be aware that this is a potential so that if it does happen particularly for us, that they will bring it up in conversation (participant 12).

v. Reflecting on changing practice to manage cognitive and behavioural change in MND

This theme describes participants' reflections about their current clinical practice and the training needs they identified that would facilitate these changes. Here, the TDF domains of *Behavioural regulation*, and *Goals* were captured.

Some participants reflected on their current clinical practice and identified benefits associated with adapting their current approaches to better support people with cognitive and behavioural changes in MND. These, specifically, related to personal attitudes and feelings for discussing cognitive and behavioural changes in MND, and how this may create a barrier for clinical discussions with families.

> I think it's going to help me as a HCP to kind of work through some of my own kind of feelings about speaking to people with cognitive and behavioural changes and how I approach that situation (Participant 09).

> I think I probably make a lot of assumptions about what I would like in that sort of position and probably make some of my clinical judgements on that, which is not ideal by any stretch of the imagination (Participant 07).

Others reflected that currently, they do not consider cognitive and behavioural changes in all pwMND and developed an awareness that this is a practice area that they could change.

> So, I think it's interesting to consider it as more of a symptom that should be looked at in every patient as opposed to just something that I see every now and then (Participant 08).

Participants expressed changes they could implement, to provide improved education to families living with MND.

> Definitely if there's an identified issue, then I usually use the MND Association handout, just the general information. But I do not. I do not provide anything in clinic. That's maybe something that I could change in terms of my practice,

> because like you say, you do get so much information that oftentimes I think people do not hear what they do not necessarily have to. Yeah, no, I only give written information if it's an already identified issue (Participant 07).

Some participants additionally identified a goal to develop skills in cognitive and behavioural changes in MND. However, they expressed a barrier to accessing additional training, believing that there were limited opportunities available to support their professional development.

> I've got to be honest, this is something which I am looking into getting as much training in as possible, I think, because it's fairly new. When I first started working with MND, it was still thought to not affect cognition at all (Participant 12).

> It's not something that comes up very regularly, because I'm always on the lookout for training. (Participant 05).

vi. Creating professional development for cognitive and behavioural changes in MND

The TDF domain of *Goals* relates to this theme and the participant's perspectives and preferences for professional development/training module development for cognitive and behavioural changes in MND are summarised. Specifically, Participants believed the provision of practical management strategies and clear communication strategies, which allow them to first learn and then educate families living with MND, would be most beneficial to support their professional development.

> …and then I think the strategies, how are we supporting them? It's sort of needs to move across the assessment, intervention, strategies type of space, I think (Participant 10).

> I think we're pretty good at recognising the signs and symptoms. It's more practical advice on how best to manage behavior that they explain, I think (Participant 05).

> And I think not just having the background information, I would want strategies and resources and things that I could actively use in clinic. So, I would want it to be more patient based as opposed to science based (Participant 08).

Additionally, participants described training would be most valuable if it was inclusive of both the lived experience of families experiencing MND and cognitive and behavioural changes, and HCPs who have had previous experience supporting families. Participants believed that including these firsthand experiences would facilitate a better understanding of the impact of cognitive and behavioural changes in MND:

> I just think it holds more meaning. And I feel like as a HCP, even though I love learning the theory and that kind of stuff, it's those moments from people with lived experience are the most impactful to me personally because it makes me kind of take a step back and apply all of that theory and go, Right, so we just learned this, which does apply to what you're saying. But you're saying you'd prefer it to be delivered from this perspective or in this manner. So, I think that kind of helps put it more into clinical practice (Participant 09).

> How do you manage this stuff? Sharing ideas, listening to experienced HCPs who've been doing it for a long time, what are the scenarios that you face, and have you dealt with them? And I think it's a lot of that practical tools that you can do (Participant 11).

Some participants identified that training programs should include information regarding the pathophysiology of cognitive and behavioural changes and be grounded in MND evidence.

In summary, several facilitators and barriers to managing cognitive and behavioural changes in MND were identified and are shown in Table 25. Specifically, participants feel confident working in their scope of practice. They know about cognitive and behavioural changes as being associated with MND, however identified gaps in their knowledge about cognitive and behavioural changes that impacts on their confidence to discuss this symptom with families. Participants identified that MDTs support both their confidence and knowledge development. Specifically, participants described consulting with colleagues to gain additional support and for gaining consensus in relation to cognitive and behavioural changes in a pwMND. Participants also identified resources burdens and an uncertainty if families with MND would want to learn about cognitive and behavioural changes as barriers to management.

Table 25.

Themes and summary of key findings related to the Theoretical Domains Framework (TDF) Domains

	THEME	TDF Domains captured within each theme	Facilitators to managing cognitive and behavioural changes identified	Barriers to managing cognitive and behavioural changes identified
i.	Knowledge of cognitive and behavioural changes in MND	Knowledge Belief about capabilities Skills Social Influences Belief about consequences	Participants know about cognitive and behavioural changes in MND; however,	Knowledge drawn from outside of an MND context. Participants identified gaps in their knowledge of cognitive and behavioural changes in MND. There is a lingering perception that cognitive and behavioural changes do not occur in MND both within community and within health care
ii.	Clinical context is both a facilitator and barrier to managing cognitive and behavioural changes in MND	Environmental context and resources Skills Social professional role & identity Belief about capabilities Belief about consequences Optimism	MDT care helps practitioners to learn and feel supported about cognitive and behavioural changes in MND Case conferences facilitate conversations about cognitive and behavioural changes in MND. Participants are confident in their skills within their scope of practice. Participants are clear on the roles and responsibilities of others in the MDT	There is unclear ownership of cognitive and behavioural changes in MND Resource burdens exist related to limited time and limited capacity for both managing cognitive and behavioural changes in MND and for professional development to develop skills.

	THEME	TDF Domains captured within each theme	Facilitators to managing cognitive and behavioural changes identified	Barriers to managing cognitive and behavioural changes identified
iii.	Approaches to managing cognitive and behavioural changes in MND	Belief about capabilities Skills Belief about consequences	Education is provided in both written and verbal formats Participants adapt management to be individualised and responsive to each families need.	Detection of cognitive and behavioural changes relies on family report, observation and/or MDT consensus Assessment and/or screening is rarely conducted
iv.	Overwhelming families living with MND	Belief about consequences Social influences		Participants believe that discussing cognitive and behavioural changes with families will be overwhelming.
v.	Reflecting on changing practice to manage cognitive and behavioural changes in MND	Behavioural regulation Goals	Participants reflected on ways to change their clinical practice related to cognitive and behavioural changes, including incorporating education into clinical discussions	

	THEME	TDF Domains captured within each theme	Facilitators to managing cognitive and behavioural changes identified	Barriers to managing cognitive and behavioural changes identified
vi.	Creating professional development for cognitive and behavioural changes in MND	Goals	Training should include i. practical management strategies for cognitive and behavioural changes grounded in in MND evidence, ii. clear communication strategies, iv. lived experiences of families with MND who have cognitive and behavioural changes, v. HCPs with expert knowledge of MND, and vi. foundational knowledge about cognitive and behavioural changes specific to MND.	

Note: (HCPs: Health care professionals; MND: Motor Neurone Disease)

Discussion

This study evaluated factors that influence clinical approaches to identifying and managing cognitive and behavioural changes in MND. We used the TDF to identify facilitators and barriers to managing cognitive and behavioural changes in MND.

Participants reported being aware that cognitive and behavioural changes are associated with MND, however, some identified gaps in their own knowledge related to the pathophysiology of cognitive decline in MND. Additionally, some participants expressed a belief that there was a lingering social perception that cognition is spared in MND. Furthermore, some participants had awareness that there are nuances to cognitive and behavioural changes in MND, with some pwMND experiencing variations of mild cognitive and/or behavioural changes, whilst others develop deficits that may impact on capacity, or result in an FTD diagnosis. Additionally, participants strongly expressed that the MND journey is individual, highly variable and functional impairments vary greatly, depending on the underlying pathophysiology and phenotype. Within MND phenotypes, there can be substantial variability in clinical presentation and rate of progression (Talman et al., 2016), which results in individualised functional impairments (Borasio & Miller, 2001). Additionally, not all pwMND will experience cognitive and behavioural changes (Lomen-Hoerth et al., 2002). Combined, these factors create several challenges to approaching cognitive and behavioural changes with pwMND and carers, including inconsistent approaches to cognitive and behavioural changes in MND, difficulty detecting cognitive and behavioural changes in MND and lack of professional development.

Challenge 1 – Inconsistent approaches to clinical management of cognitive and behavioural changes in MND

This study highlighted that providing information about cognitive and behavioural changes in MND occurs inconsistently. For example, some participants felt unsure if families would want

to know about cognitive and behavioural changes. In this instance, some participants expressed concerns that this information would add to the overwhelm that families living with MND are already facing. Given the variability and individuality of MND, participants described enacting responsive practice, relative to their discipline, to provide symptomatic relief and optimise quality of life.

Challenge 2 – Difficulty detecting cognitive and behavioural changes in MND

A further challenge highlighted in this study relates to the detection of cognitive and behavioural changes in MND. Within usual practice, participants described considering the complexity of the severe functional impacts and the immediate needs of each individual pwMND, as they support them and their family to manage overt symptoms. This approach aligns with the evidence-based practice of person-centred care (Australian Commission on Safety and Quality in Health Care, 2023), where a person's individual needs are considered and management decisions are made in partnership with families. However, as MND often results in severe functional deficits, related to impaired mobility and impaired speech, coexisting cognitive and behavioural changes in MND can be difficult to detect. The findings of this study show that participants mostly rely on observation and clinical judgement to alert them to potential changes to cognition and behaviour. In addition to clinical observations, participants described seeking support from MDT members to reach consensus on their suspicion of cognitive and behavioural changes. However, detection of cognitive decline via observation in a primary care setting, via clinical judgement alone, has been shown to be unreliable (Mitchell et al., 2011).

Specifically, identification of mild cognitive impairment (MCI), as described in the dementia literature, is challenging (Kaduszkiewicz et al., 2010). Detection of MCI from clinical judgment alone is poor, with accurate detection rates occurring in less than half of MCI cases (Mitchell et al., 2011). It is documented that often, people who experience changes to their thinking or memory, may not actively seek help from their general practitioner (Mitchell

et al., 2011). In addition, pwMND often present to an MND HCP when they have developed motor deficits and/or dysarthria, meaning the clinical signs of cognitive decline related to physical acts or communication may be masked by other symptoms of MND. This further complicates the detection of cognitive and behavioural changes in MND. Additionally, as these are often specialist clinics, there may be no pre-disease patient-doctor relationship for HCPs to draw on as a point of reference to help them identify changes to cognition or behaviour in individual's. Furthermore, the newly diagnosed pwMND and their family are also likely to be experiencing the devastating psychological impact of receiving a terminal diagnosis. As a consequence, HCPs may link cognitive and behavioural symptoms to psychosocial factors. For example, apathy may be mistaken for depression, because clinically apathy and depression share similar characteristics. However, it is important to differentiate apathy from depression because apathy in MND is documented to predict poorer prognosis (Caga et al., 2016), negatively impact on a pwMND's quality of life and increase caregiver burden (Caga et al., 2018). In fact, apathy in MND is also associated with increased rates of depression (Caga et al., 2018).

Apathy has been shown to be the most common behavioural symptom in MND (Caga et al., 2018). However, interestingly, when participants in this study described cognitive and behavioural changes in MND, only one described apathy. Apathy is a commonly occurring dementia-related symptom, documented to occur in approximately half of people with mild Alzheimer's disease (Tagariello et al., 2009). Apathy may present as decreased interest, impaired emotional expression and a reduced ability to initiate tasks that individuals have the physical capability to do. As a consequence apathy is often mistaken for depression (Lanctôt et al., 2023; Tagariello et al., 2009). Notably, depression and apathy can co-occur, and they share similar clinical presentations, so differential diagnosis is difficult. However, it is important to do so, as in other neurodegenerative diseases such as Parkinson's Disease and FTD, the rate of progression and severity of apathy is linked to disease progression (Lanctôt et al., 2023). Additionally, antidepressant medications used to treat depression have

been associated with increased rates of apathy in people with dementia (Lanctôt et al., 2023). This is concerning as antidepressant medications have been described as being a treatment for cognitive and behavioural changes in MND (Chapter 6) and it is unclear if this treatment approach also poses as a risk for increased apathy in MND. Taken together, these factors highlight the importance of differentiating apathy, associated with cognitive and behavioural changes in MND, from depression. To do this, MND care teams need to participate in clinical conversations which support a pwMND to self-report their experience, as well as conduct cognitive and behavioural assessments which include assessment of executive functions. Additionally, assessments of psychosocial functioning should be conducted, to support accurate identification of apathy and depression.

Notably, within this study, no participant described asking a pwMND directly if they had become aware of any changes to their thinking and behaviour. Rather, some participants expressed clinical conversations about cognitive and behavioural changes occurring only if the topic is raised by families. However, this approach appears suboptimal, as members of the community seek guidance and information from their HCPs and may lack awareness of what they need to understand about MND, what to look out for and what to ask about. Further, there is a lingering community perception that cognition is spared in MND, meaning pwMND or carers may not relate their experiences of changes to thinking and/or behaviour as being part of MND. Consequently, HCPs play a vital role in providing education to families experiencing MND to enable both pwMND and their carers to identify these changes should they arise. However, it remains unclear where the ownership of addressing cognitive and behavioural changes in MND with families belongs.

Challenge 3 – Unclear ownership of addressing cognitive and behavioural changes in MND

Some participants described that addressing cognitive and behavioural changes may fall outside of their own scope of practice, or they were unclear about who takes ownership of addressing this with families. Overall, participants reasoned that the professional

responsibility to identify cognitive and behavioural changes would typically sit within the role of a neurologist, neuropsychologist or occupational therapist. Further, some participants described making referrals to other disciplines, or an MND service, as a way to manage cognitive and behavioural changes in MND. However, participants also reported that the clinical care model that they work within may not have access to these disciplines, or that a pwMND would experience long wait times for a service. As pwMND often have rapidly progressing neurodegeneration, the need to address cognitive and behavioural changes quickly becomes increasingly more critical. It has previously been recommended that cognitive and behavioural changes in MND are considered in clinical management more broadly (Goldstein & Abrahams, 2013), and that any HCP in an MND MDT care team can conduct assessments for cognitive and behavioural changes (Gray & Abrahams, 2022). Consequently, it is suggested that HCPs acquire ongoing knowledge of a pwMND's cognitive status when providing management within their scope of practice. This represents the third challenge for managing cognitive and behavioural changes in MND outlined in this study.

Challenge 4 – Lack of professional development

Participants reported being unable to fully support pwMND as they would like to and being unable to seek out professional development opportunities to increase their knowledge of cognitive and behavioural changes in MND due to resource constraints. Specifically, participants expressed that they felt under-resourced with time, staffing and funding. In order to support HCPs to develop their knowledge and skills, it is important to provide staff with dedicated time for professional development and opportunities to participate in training about cognitive and behavioural changes in MND. Workforce upskilling in this space is particularly important, because participants identified gaps in their knowledge related to cognitive and behavioural changes in MND and described that this lack of knowledge undermined their confidence in discussing this with families.

When MND HCPs or MDTs are under-resourced and operate on restrictive consultation schedules, HCPs may prioritise more tangible medical facets of care. As such they may not have the capacity to identify, and subsequently manage, cognitive and behavioural changes for these reasons. However, upskilling all disciplines who work with an MND caseload to identify and assess for cognitive and behavioural changes would potentially have several benefits. First, this may provide families with MND with a more efficient service and secondly, it may go towards alleviating the feelings of discomfort described by participants who may not have the appropriate skills to assess and manage cognitive and behavioural changes in MND.

Possible solutions

One possible solution to the challenges outlined in this study may be to develop clearer clinical frameworks for cognitive and behavioural changes in MND. Clinical frameworks are used to describe guiding principles and elements of care with the aim of delivering the best outcomes for patients (Aged Care Quality and Safety Commission, 2019). The development of a clinical framework for cognitive and behavioural changes in MND would enable HCPs to feel confident, first in discussing cognitive and behavioural changes with families and second, in enacting processes to identify and support families to manage cognitive and behavioural changes. A clinical framework would also enable conversations about cognitive and behavioural changes to become part of routine clinical practice. An important step towards the development of clinical frameworks would be to first learn from both pwMND and carers living with MND, to understand their preferences in relation to cognitive and behavioural changes.

For instance, it is important to first establish whether pwMND and carers want to receive information about cognitive and behavioural changes and if so, when in the disease journey should this information be provided. Additionally, it would be important to learn families' preferences for how, and who they would like to receive this information from. By designing

future studies that employ co-design methods, together with families with lived experience of MND, may enable a better understanding of optimal cognitive assessment and screening processes. Notably, recent studies have found that implementing regular screening practices result in positive outcomes for both pwMND and carers (Gray & Abrahams, 2022). To this end, further evaluation of pwMND and carers preferences for cognitive assessments would be valuable. Overall, working with families to understand their preferences would enable a better understanding of how information about cognitive and behavioural changes is gathered and shared with families, while considering the abovementioned challenges and the overwhelming nature. Through this, it is important to recognise that cognitive and behavioural changes are perceived individually, which in turn may impact on how much they affect an individual's functioning.

Given the individual and progressive nature of MND, training materials are needed that allow for and facilitate development of the HCPs' ability to generate and then internally and cyclically test their clinical reasoning. This approach to HCP training will enable HCPs to adjust and review strategies to devise ongoing management plans that are dynamic and tailored to an individual's needs as MND progresses. Throughout, it will be important to consider individual circumstances and preferences, given the high variability of MND presentations, combined with pwMND's unique beliefs and wishes. Furthermore, these resources must be inclusive of strategies that are aimed at supporting carers to reduce caregiver burden.

The outcomes of these future studies have the protentional to inform and advance professional development and training modules that support HCPs to upskill in this area of MND clinical practice. As was highlighted by participants in this study, future professional development and clinical resources should also be inclusive of clear, practical and evidence-based management strategies and support clinical reasoning.

Limitations

A limitation of this study was the sample size, which may restrict the perspectives and limit transferability of the findings to broader populations. Additionally, it is acknowledged that most participants were located in Australia and the UK, and, as such, the results may not be reflective of care practices in other countries.

Conclusion

MND is highly variable across and within each individual, depending on disease onset and rate of progression. Subsequently care practices need to be tailored and responsive. The functional impairment of having cognitive and behavioural changes within the context of person's individual presentation and needs to be considered to guide management.

The previous chapter showed that HCPs who support pwMND have an understanding that cognitive and behavioural changes are associated with MND. However, the clinical approaches to discussing this with pwMND and carers, identifying changes and subsequently managing cognitive and behavioural changes, are highly variable. HCPs felt strongly that each pwMND has a unique and individual experience of the disease. Some held the view that pwMND want to learn about cognitive and behavioural changes in MND, whereas others described that pwMND may be overwhelmed. In a described effort to protect their patients, the topic of cognitive and behavioural changes appears to feature less frequently in clinical discussions about the disease. Therefore, it remains unclear how pwMND and carers experiencing MND learn about, experience and manage cognitive and behavioural changes.

The following chapter describes the findings of a qualitative study, in which semi-structured interviews were conducted with both pwMND and carers. The study is described in manuscript format, with the intention of submitting this manuscript to the International Journal of Speech-Language Pathology. The aim of this study was twofold. The first aim was to better understand how families living with MND experience the symptoms of dysphagia and cognitive and behavioural changes. The second aim of this study was to explore if cognitive and behavioural changes in MND impact on managing dysphagia in the home. Participants were asked questions about their experiences of both dysphagia and cognitive and behavioural changes, as well as their preferences for receiving care.

CHAPTER 8: FAMILIES' EXPERIENCES AND PREFERENCES FOR MANAGEMENT OF DYSPHAGIA AND COGNITIVE AND BEHAVIOURAL CHANGES IN MND

Introduction

Almost all pwMND will develop dysphagia over the course of disease progression (MND Australia, 2021a). Dysphagia in MND is associated with an increased risk of respiratory difficulties, such as choking and aspiration pneumonia (MND Australia, 2021). In addition, dysphagia in MND is associated with dehydration, malnutrition and a decreased quality of life (Lisiecka et al., 2021; Waito et al., 2017). Consequently, speech pathologists are involved in MND MDTs, to support pwMND to manage their swallowing difficulties, and to continue to eat safely via oral route. These management plans may consist of education about food and liquids that may pose greater swallowing risk, diet modifications and/or behavioural interventions. To maximise swallowing safety, a speech pathologist may recommend diet modifications that involve altering food and liquid consistencies, as well as recommending a pwMND avoids eating certain foods that are assessed to be of higher risk when swallowing. In addition, behavioural interventions to support a pwMND's swallowing efficiency include motor behavioural techniques, such as postural adjustments or swallowing manoeuvers. However, receiving education about, and subsequently implementing management strategies for dysphagia within the home, requires a pwMND to cognitively engage. Specifically, a pwMND needs to understand the swallowing problem and the associated risks, understand the management strategy, and enact and adhere to suggested management plans. There is an additional layer of complexity in managing dysphagia in MND, however, as cognitive and behavioural changes are also known to be associated with MND (Motor Neurone Disease Association, 2022).

It is estimated that up to 50% of pwMND experience cognitive and behavioural changes, which can impact on thinking and behaviour (Clarke & Levine, 2011; Goldstein & Abrahams,

2013; Neary et al., 2000b; Phukan et al., 2007). However, as shown in previous studies (Francis et al., 2021), the interaction of cognitive and behavioural changes and dysphagia in MND is unknown. As described in Chapter 5, approaches to the assessment and management of cognitive and behavioural changes in MND are highly variable (Francis et al., 2023) and in some cases, cognitive changes may be missed altogether. However, evaluation of cognitive and behavioural changes in MND is important, as these changes are linked to negative outcomes for both the pwMND and their carer (Radakovic et al., 2024). Further, there are implications for how other symptoms of the disease are managed in the presence of cognitive and behavioural changes. For example, cognitive and behavioural changes have been reported to reduce treatment uptake related to non-invasive ventilation and enteral feeding procedures frequently used in MND management (N. H. Martin et al., 2014). To add to this challenge, it is also currently unclear how families with MND experience cognitive and behavioural changes. Specifically, it is not known how families prefer to receive information about cognitive and behavioural changes, and how they would prefer subsequent management to occur, if at all.

Individual preferences for assessment and management of cognitive and behavioural changes are important to understand, as these have been linked to poorer outcomes for both the pwMND, their carers, and wider family units (Oseland et al., 2023). Specifically, behavioural changes in MND have been shown to negatively impact on a pwMND's quality of life (Radakovic et al., 2024) and are linked with increased strain, burden and distress for carers (de Wit et al., 2018; Pagnini et al., 2010). However, as outlined in previous chapters of this thesis, there are substantial inconsistencies in care approaches for cognitive and behavioural changes in MND. For example, inconsistencies in assessment practices may mean that cognitive and behavioural changes are not always identified in pwMND. Further, inconsistencies in management of cognitive and behavioural changes in MND may mean that families do not receive education about these changes as being associated with MND. As discussed in Chapter 7, pwMND may not be explicitly asked if they have experienced

changes to their thinking or behaviour. HCPs may instead rely on clinical observation or carer report to identify cognitive and behavioural changes. However, clinical observations of mild-moderate cognitive changes as an identification method have been found to be unreliable (Kaduszkiewicz et al., 2010) and identification of cognitive and behavioural changes exclusively through carer report or observation may also be unreliable, coincidental or incomplete. Additionally, as shown in Chapter 6 and 7, educational discussions between HCPs and pwMND and carers about cognitive and behavioural changes are inconsistently conducted.

As families may not be aware that cognitive and behavioural changes are associated with MND, this limits their ability to link experienced or observed changes to something that could be reported to, and discussed with, their healthcare team. For example, carers may mistake behavioural changes as changes to a pwMND's mood and as a consequence, receive changes in a pwMND's behaviour in a negative way. Consequently, carers may feel offended or upset by behaviours that are not consistent with the pwMND' personality prior to their diagnosis (Oseland et al., 2023). As a consequence, relationship breakdowns and increased burden within family relationships may occur. Additionally, as pwMND and carers are unaware that these changes are associated with the disease, they potentially will not report this back to their health care team. Therefore, undertaking educational discussions about cognitive and behavioural changes early in disease progression may help to alleviate these misunderstandings and equip pwMND and carers with the information they need to be aware that cognitive and behavioural changes can occur.

Early educational discussions may also facilitate regular and ongoing check in conversations between families and their HCPs about cognitive and behavioural changes, as well as supporting families to make informed decisions about proceeding or not proceeding with cognitive assessments. Further, learning of a family's readiness to receive education and their preferences for assessment and subsequent management, may also alleviate

additional burdens as being associated with cognitive and behavioural changes. Additionally, this knowledge may help to improve clinical pathways and support the development of literature to guide HCPs through clinical practices related to cognitive and behavioural change in MND to support both the pwMND and the carer.

To this end, we conducted a qualitative study using semi-structured interviews with pwMND and their carers and aimed to learn about their experiences of both cognitive and behavioural changes and dysphagia. Additionally, we aimed to understand how pwMND and their carers prefer to receive information on, undergo assessment for and subsequently manage, cognitive and behavioural changes and dysphagia. To do this, we employed the TDF to facilitate understanding and to identify influences on capability, beliefs and motivational factors that underpin behaviour.

The TDF framework (Table 26) was originally designed for use to identify barriers and facilitators amongst health care settings (Atkins et al., 2017). However, it has also been shown to be an effective theoretical model to apply to studies of lived experience of disease (Baay et al., 2019; McGowan et al., 2020). As an integrative framework, the TDF consists of 84 constructs and 14 domains of theory-based explanations of behaviour. This framework allows data analysis of both the personal and environmental contexts to determine influence on behaviours (Pagnini et al., 2010). We applied this framework to answer the following research question:

How do pwMND & carers experience the symptoms of dysphagia and cognitive and behavioural changes in MND, and what are their preferences for management?

Table 26.

The theoretical domains framework domains with corresponding descriptor

Domain and definition	Constructs
1. Knowledge (An awareness of the existence of something)	Knowledge (including knowledge of condition/scientific rationale) Procedural knowledge Knowledge of task environment
2. Skills (An ability or proficiency acquired through practice)	Skills Skills development Competence Ability Interpersonal skills Practice Skill assessment

Domain and definition	Constructs
3. Social/professional role and identity (A coherent set of behaviours and displayed personal qualities of an individual in a social or work setting)	Professional identity Professional role Social identity Identity Professional boundaries Professional confidence Group identity Leadership Organisational commitment
4. Beliefs about capabilities (Acceptance of the truth, reality or validity about an ability, talent or facility that a person can put to constructive use)	Self-confidence Perceived competence Self-efficacy Perceived behavioural control Beliefs Self-esteem Empowerment Professional confidence
5. Optimism (The confidence that things will happen for the best or that desired goals will be attained)	Optimism Pessimism Unrealistic optimism Identity

Domain and definition	Constructs
6. Beliefs about Consequences (Acceptance of the truth, reality, or validity about outcomes of a behaviour in a given situation)	Beliefs Outcome expectancies Characteristics of outcome expectancies Anticipated regret Consequents
7. Reinforcement (Increasing the probability of a response by arranging a dependent relationship, or contingency, between the response and a given stimulus)	Rewards (proximal/distal, valued/not valued, probable/improbable) Incentives Punishment Consequents Reinforcement Contingencies Sanctions
8. Intentions (A conscious decision to perform a behaviour or a resolve to act in a certain way)	Stability of intentions Stages of change model Transtheoretical model and stages of change

Domain and definition	Constructs
9. Goals (Mental representations of outcomes or end states that an individual wants to achieve)	Goals (distal/proximal) Goal priority Goal/target setting Goals (autonomous/controlled) Action planning Implementation intention
10. Memory, attention and decision processes (The ability to retain information, focus selectively on aspects of the environment and choose between two or more alternatives)	Memory Attention Attention control Decision making Cognitive overload/tiredness
11. Environmental context and resources (Any circumstance of a person's situation or environment that discourages or encourages the development of skills and abilities, independence, social competence and adaptive behaviour)	Environmental stressors Resources/material resources Organisational culture/climate Salient events/critical incidents Person × environment interaction Barriers and facilitators

Domain and definition	Constructs
12. Social influences (Those interpersonal processes that can cause individuals to change their thoughts, feelings, or behaviours)	Social pressure Social norms Group conformity Social comparisons Group norms Social support Power Intergroup conflict Alienation Group identity Modelling
13. Emotion (A complex reaction pattern, involving experiential, behavioural, and physiological elements, by which the individual attempts to deal with a personally significant matter or event)	Fear Anxiety Affect Stress Depression Positive/negative affect Burn-out
14. Behavioural regulation (Anything aimed at managing or changing objectively observed or measured actions)	Self-monitoring Breaking habit Action planning

Methods

We employed a descriptive qualitative study design, consisting of semi-structured interviews, to explore how pwMND and their carers experience both cognitive and behavioural changes and dysphagia in MND. Ethics approval (Proposal #4660) was gained from Flinders University (Appendix 01).

Two topic guides were created, one specific for a pwMND and the other specific for a carer. The interview topic guides (Appendix 11) were informed by both the existing literature and the previous studies that contribute to this program of research, namely the literature review in Chapter 3 and the HCP survey (Chapter 6) and interviews (Chapter 7). Interview questions were mostly open-ended, and designed to elicit information about the interviewee's knowledge, experience, beliefs and preferences related to the research question. The topic guides were set out in two sections. The first section explored the experiences of dysphagia in MND, whereas the second section explored cognitive and behavioural changes in MND. These sections allowed for the questions to flow in a logical sequence and aimed to support the interviewees' thinking on each topic. The draft questions were refined via an iterative process amongst the broader research team. Once finalised, an amendment to the ethics application was submitted with the finalised topic guides for approval (Appendix 1, 11). As mentioned above, the topic guide for this study was informed by the findings of the previous studies that contributed to this body of work. This meant that the interview questions were finalised after the findings of the previous studies were complete.

Participants and recruitment

Any person with a confirmed or probable diagnosis of MND and carers of a pwMND were eligible to participate in this study. The role of carer was defined as any person who fulfilled the role of providing daily care to a pwMND. This included for example, a person's spouse, any family member, or a professional support worker, if they were the pwMND's only or primary carer. A research flyer (Appendix 9) was distributed via social media platforms and newsletters, including the MND Association in the UK, MND services in Australia, as well as personal social media platforms of the research team. Additionally, the MND Association in the UK disseminated the details of the study within their newsletter (Appendix 15). Any pwMND or carers were invited to participate and advised to contact the researcher via email. Upon receipt of an email enquiry, participants were forwarded the participant information sheet and consent form (PICF) via email (Appendix 12,13). Each participant was provided with the equivalent of an AUD$40 honorarium in the form of a gift card.

Study Design

This study was grounded in constructivism and interpretivism paradigms (Alharahsheh & Pius, 2020; Charmaz, 2006, 2008), which asserts that people draw meaning from their own experiences. Therefore, knowledge is socially and actively constructed, with learning and meaning making occurring through an individual's interaction with others and with their environment. Within qualitative research, the constructivist interpretivism approach centres on exploring perspectives and experiences to gain a deeper understanding of the phenomenon being investigated (Adom et al., 2016).

Procedures and data collection

Consented participants took part in semi-structured interviews, conducted throughout 2023. Interviews were either via telephone or online video meetings using either Microsoft Teams or Zoom, depending on which was most accessible for the participant. With verbal consent, each interview was recorded. Participants who were participating as a pair of a pwMND and carer were given the option of completing the interview together or separately in

consideration of the pwMND's communication difficulties. For example, if a pwMND was able to communicate verbally, then an interview with their carer was conducted separately. When the interviews were completed in a dyad (with pwMND and their carer), the interviewee directed specific questions to either member to provide their individual answer (as guided by the separate interview guides)

Analysis

All video and voice recordings were transcribed verbatim, removing names and locations to protect participant anonymity. The method for creating the transcripts varied depending on the interview source, and the level of communication difficulty the pwMND had. Specifically, transcripts were created either via an external transcription service, or voice to text functionality within Microsoft Teams or Microsoft Word. Following transcription creation, all transcripts were checked against the original interview for accurateness.

Initially, the candidate reviewed the interview transcripts, undergoing a process of data immersion to familiarise themselves with these data. Further, memo taking was conducted at this stage, as a summary of thinking and to summarise the setting, feelings and general thoughts of the candidate related to the research question. Data were coded and analysis was completed using NVivo software. First, as a way to organise the data, initial coding was conducted deductively to the domains of the TDF (Atkins et al., 2017; McGowan et al., 2020; McSherry et al., 2012) and followed a constant comparative approach where the identified themes were represented across the interview transcripts and initial coding to ensure the themes were grounded in the data. This enhanced rigour by ensuring that the deductive process had not fractured the themes and interpretation from the intentions of the interview. Hence, during the initial coding process, annotation and memo creation was conducted to support thinking and for later sense checking within the broader research team.

Once the first round of deductive coding was complete, the candidate undertook iterative inductive analysis, first fracturing the data to create subcodes that sat under each of the TDF

domains as shown in Figure 30. Then these subcodes were reviewed to identify categories and patterns within the data.

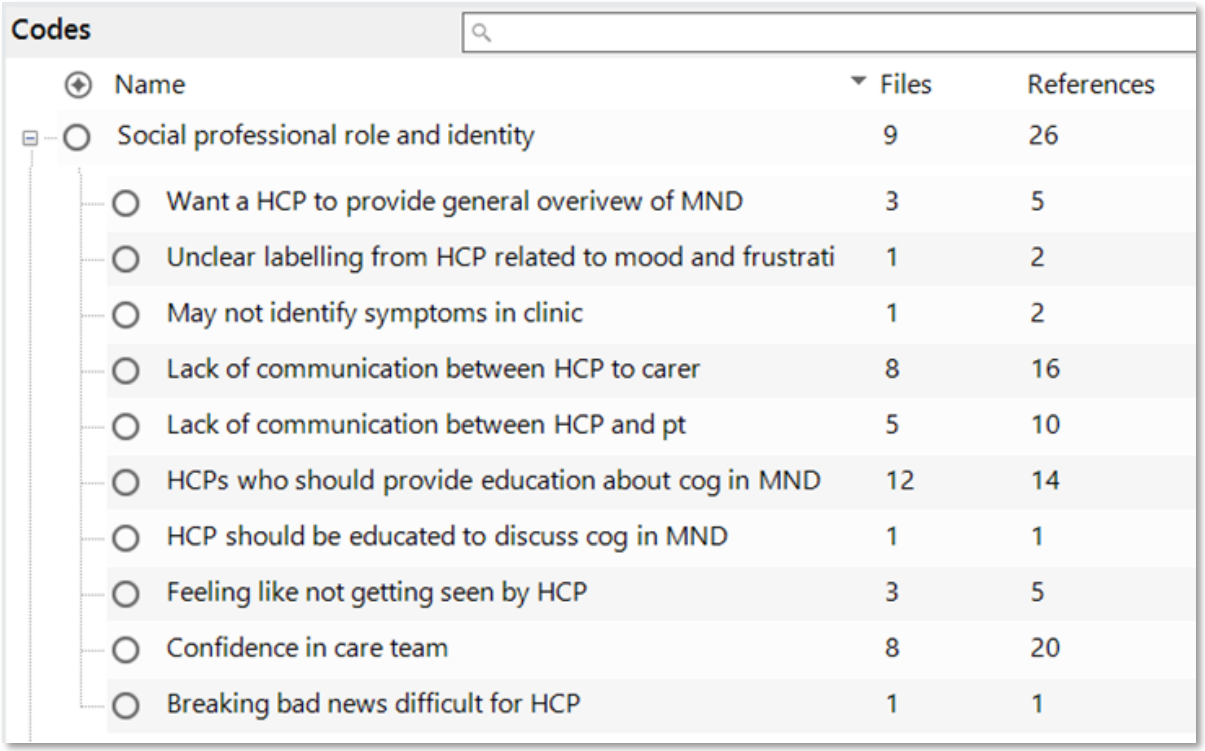


Figure 30: Example of data fracturing within the TDF domain of *Social professional role and identity. (TDF: Theoretical Domains Framework)*

Following this, a process of thematic analysis was undertaken. Thematic analysis allows for flexibility in data analysis and may be used for both inductive and theory driven deductive analysis (Braun & Clarke, 2017). Further, it acknowledges the importance of identifying the recurrence of patterns within the data, while at the same time also constructing meaning and meaningfulness throughout the coding process (Braun & Clarke, 2006). Consequently, all subcodes under each domain of the TDF were inductively analysed to search for themes. Finally, consultation with the broader research team took place, to define and finalise these themes.

Findings

Participants

Twenty-four individuals agreed to participate in this research study. As shown in Table 27 and Table 28, participants were located in Australia (n=14), the UK (n=9) and Kenya (n=1). The average length of each interview was approximately 50 minutes. Of the participants, twelve were pwMND (M=11, F=1) with an average age of 66.5 years (range 51yrs to 81yrs). In addition, there were twelve carers (M=3, F=9), with an average age of 60.2 years (range 42yrs to 81yrs). In total, there were fifteen interviews conducted. Five pwMND and five coMND were in a spousal relationship, and their interviews were conducted asynchronously, three pwMND and three coMND were in a spousal relationship and were interviewed synchronously. Within the group of carers, four participated individually and three of these carers were individuals whose family member had passed away, and they provided reflective answers about their past experiences with MND. Further, one participant was a professional support worker and primary carer of a pwMND. As the inclusion criteria stated any carer of a pwMND could participate, this person met the eligibility criteria. Additionally, some participants were no longer able to communicate verbally. Therefore, one participant used an AAC device (text to talk) to answer the interview questions, and three participants, who were anarthric received the interview guide via email and provided their responses to the questions in writing.

Table 27.

Participants demographics (pwMND)

Participant Code	M/F	Location	Age	Type of MND	Diagnosed
PWMND001	M	SA (Semi-rural)	75	PLS	2013
PWMND002	M	SA	81	ALS	2022
PWMND003	M	SA	59	ALS	2015
PWMND004	M	SA	70	Initial ALS/ Revised to PLS	2018
PWMND005	M	VIC	65	ALS	2022
PWMND006	M	SA (Rural)	68	ALS	2022
PWMND007	M	UK	64	ALS Bulbar onset	2023
PWMND008	F	UK	67	PLS	2014
PWMND009	M	UK	51	ALS Upper limb onset	2017
PWMND010	M	UK	61	Suspected PLS	2020
PWMND011	M	UK	66	ALS Bulbar onset	2023
PWMND012	M	UK	70	Unknown	2021

Note (pwMND: person with Motor Neurone Disease)

Table 28.

Participant demographics coMND

Participant code	M/F	LOCATION	AGE	CARER OF
coMND001	F	SA (Semi-rural)	69	Spouse
coMND002	F	SA	56	Spouse
coMND003	F	SA	81	Spouse
coMND004	F	SA	47	Client / SW
coMND005	F	VIC (Rural)	64	Spouse
coMND006	F	SA (Rural)	Not given	Spouse
coMND007	F	NSW	61	Spouse voice
coMND008	F	NSW	50	Mother
coMND009	M	Nairobi (Kenya)	62	Brother
coMND010	M	UK	61	Wife
coMND011	F	UK	42	Husband
coMND012	M	UK	62	Wife

Note: (coMND: carer of person with Motor Neurone Disease)

Themes

To support elucidation of the following themes, deductive/inductive analysis was completed, and the following six themes (Figure 31) were derived from the interview data:

i. *What I have experienced*

ii. *What I have experienced related to my care*

iii. *How this affects me*

iv. *If I had known, it might have been different*

v. *What I do to manage*

vi. *I want care that is relational, individualised, informative and empowering*

The domains of the TDF, as they relate to each theme, are described to aid understanding of the lived experience of dysphagia and cognitive and behavioural changes in MND, the impact of dysphagia and cognitive and behavioural changes, and the person's preference for health care management of these.

The findings shown in the themes; *What I have experienced, What I have experienced in relation to my care* and *How this affects me,* describe the experiences of pwMND and their carers in relation to cognitive and behavioural changes and dysphagia more broadly (Figure 31). Within each theme, each relevant TDF domain is described to provide greater context to this program of research and the findings will be further illustrated with quotes. The first three themes answer the first part of the research question related to experience, and the impact of these experiences. Further, as also shown in Figure 31, there are additional subthemes that relate to the lived experiences of either the pwMND, or the carer, specific to either dysphagia or cognitive and behavioural changes. Then, in the themes of *What I do to manage,* and *I want care that is relational, individualised, informative and empowering,* the included findings are directly linked to the second part of the research question which discusses personal preferences for care and management of dysphagia and cognitive and behavioural changes. Quotes are also included here to illustrate these findings.

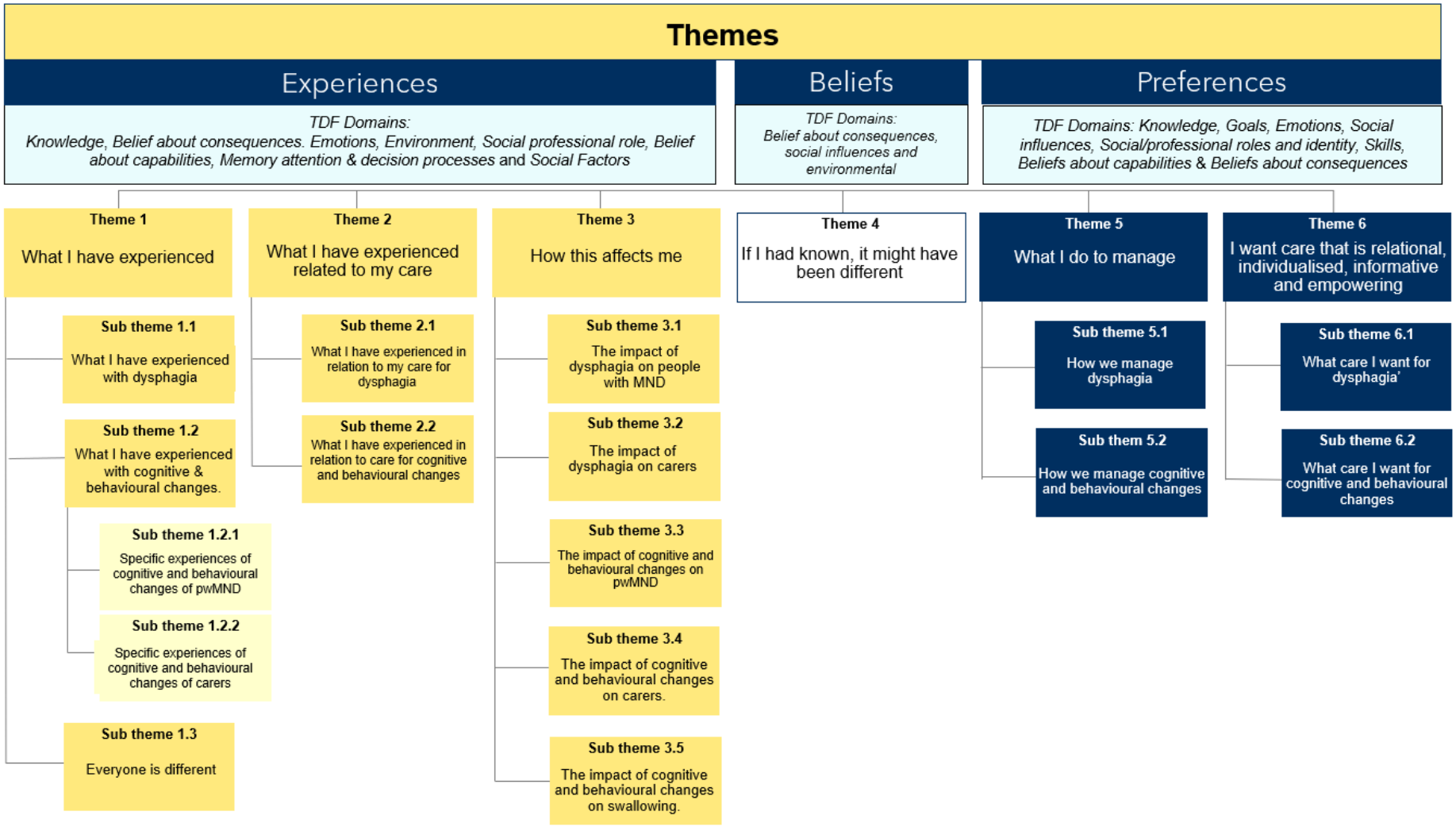


Figure 31. Themes and subthemes. Chapter 8
(MND: Motor Neurone Disease; pwMND: People with MND; TDF: Theoretical Domains Framework). **Image created by thesis author.**

Theme 1. What I have experienced

This theme describes the experiences of pwMND and carers. The term ‘experience’ for the purpose of this theme encompasses participant’s feelings, encounters, past actions or observations. Collectively, within this theme, the TDF domains of *Knowledge*, *Belief about consequences, Emotion, Environmental context and resources* and *Social Influences* play a role. The data are presented in three subthemes.

1.1, What I have experienced with dysphagia

1.2, What I have experienced with cognitive and behavioural changes’ with the subthemes

1.2.1, Specific experiences of pwMND
1.2.2, Specific experiences of carers

1.3, Everyone is different.

Sub theme 1.1 What I have experienced with dysphagia

All participants had knowledge that dysphagia is associated with MND. The participants with MND, who have also experienced dysphagia, described a feeling of ‘choking’, or of ‘things becoming stuck’ in their throat when trying to swallow. In addition, ‘coughing’ on food, fluids or secretions was frequently reported as an experience related to dysphagia in MND. Further, pwMND described that dysphagia requires them to be consciously aware of the foods they choose and avoid. They also reported feeling that they must remain attentive when eating and drinking to maintain airway protection. Eating was described by some participants as getting harder.

> Slowing down when you're enjoying something you like to eat is hard, but you have to concentrate on the fact that even though you're enjoying what you are eating, you still need to be careful. Like eating chips, I like hot chips, but normally I'd eat them fairly quick. But nowadays, even chips, I've got to be careful (pwMND002).

> Certainly, some foods I avoid completely like steak and chunky food. I tend to eat easy stuff that goes down easily, so it's certainly gotten harder, but I'm fairly particular in

> what I eat now because I can tend to get things stuck in my throat and tend to cough a lot (pwMND003).

However, the depth of knowledge which some participants described in relation to their swallowing function or dysphagia more broadly was at times limited, with participants expressing that they knew their swallowing function was impaired but were less clear on how or why it had changed:

> Um, just on the fact that it was part of my motor neurone disease and that your throat would close rather than everything else that went on with it (coMND008).

> Well, all I know is it's difficult to swallow and I've had the PEG tube put in because they said time will come when you cannot swallow (pwMND002).

Additionally, some participants described gaps in their knowledge related to dysphagia. One carer described that they did not know that aspiration pneumonia is a consequence of dysphagia and is an associated risk factor of dysphagia in MND. Two other participants described emotions of uncertainty post a percutaneous endoscopic gastrostomy (PEG) procedure. These participants described limitations in their knowledge of how to manage their PEG, specifically related to cleaning, and expressed concerns about infection.

> Probably the PEG was the thing we had the least instruction about. The maintenance of the PEG wound site…we're still a bit confused how best to do it (coMND006).

Lastly, some participants reported that they had not yet experienced changes to their/their pwMND's swallowing, however, described knowledge that dysphagia was associated with MND. These participants also reported that they were linked in with a speech pathology service to support them when the need arises.

Sub theme 1.2 What I have experienced with cognitive and behavioural changes

The participants' experience of cognitive and behavioural changes related to MND was described in a different way compared to how participants described dysphagia. Specifically, participants were more knowledgeable about swallowing function. Consequently, many participants, both pwMND and carers, reported gaps in their knowledge related to cognitive and behavioural changes associated with the disease. For example, some participants reported that they did not know that cognitive and behavioural changes are associated with MND and revealed that they only learnt about this through participating in this research.[11] Generally, participants expressed that MND was more frequently talked about as impacting mobility and perceived a lack of understanding within the broader community that cognitive and behavioural changes may be associated with MND. Additionally, some participants felt that there were social repercussions associated with cognitive decline and described that, in their experience, cognition was not freely talked about or adequately addressed in the MND community, or the community more broadly. Some participants expressed their view that there is a societal stigma associated with talking about cognitive changes.

> It does not get talked about enough. It really does not (coMND001).

> I think there's still a lot of stigma about that and a lot of unwillingness to think that there may be an issue like that, or that if there is an issue like that, then it says something fundamentally bad about you (coMND012).

Sub theme 1.2.1. Specific experiences of cognitive and behavioural changes of pwMND

Approximately half of the participants with MND had knowledge that they have experienced changes to their cognition and behaviour, with the following characteristics described; impaired decision making, rigidity in thinking, increased stress and anxiety, apathy and a reduced ability to show empathy.

[11] Note: In this case, the candidate provided these participants (n=6) with verbal education about cognitive and behavioural changes in MND, and after concluding the interview posted out an MND Association resource to support their understanding).

> Well, it's my way or the highway and I will not let anyone change my mind (pwMND001).

> Like the empathy side. I'm definitely, I even know when it's happening, but I've got no control of it (pwMND008).

> That is the biggest change (pwMND006). *(Note: pwMND is referring to feelings of increased stress related to decision-making)*

> Now that I'm experiencing it, I'm far more aware of this problem. I was okay probably, again, up to 18 months ago. But I find now that my thinking and if I've got to do something complicated, it's quite hard. So, I certainly know and experience the problem. So, I did not know much about it in the early stages, but now that I'm experiencing it, I'm becoming more aware of the problem (pwMND003).

> I think the thing is with MND, your concentration levels are depleted (pwMND003).

Additionally, one participant described feelings of apathy in relation to eating.

> Yeah. There are some things that nowadays I just think I cannot be bothered. Even sometimes *** will ask me what I want to eat and sometimes even when I'm hungry, trying to think of what I want to eat, sometimes I just say, I'm hungry, but I cannot think of what I want and cannot be bothered trying to think of what I want (pwMND003).

Sub theme 1.2.2. Specific experiences of cognitive and behavioural changes of carers

Over half of the participants who cared for someone with MND also described knowledge of cognitive and behavioural changes, and shared stories of their own personal experience. Presentation of cognitive and behavioural changes were described by carers as: an inability to process information, impaired reasoning, impaired problem-solving and decision-making, reduced ability to pay attention, personality changes, increased stress and agitation, decreased empathy,

rigidity and fixed thinking. To highlight the experiences of carers and provide broader context and convey emotions, the candidate has chosen to include several longer quotes from carers.

> He does understand it, but he does not follow through on what he understands. He's laughing at me, but it's very true. It's almost like it does not stick in the head, and if he just wants something, that's it. He has it. Yeah… It is not the person that's changed. ***'s still there, but he's not acting the way he used to. He does not have the empathy that he had before. They lose their empathy (coMND001).

> But one thing I did definitely notice with him is his reasoning. His reasoning changed a lot…And I just remember the first time watching him, he did not know how to deal with it. He's standing there. He was trying to think, how do I deal with this? And I remember thinking, He can't. He has not got the capacity to think about this. And this is normally something that he would go, yep. Let's do this, this and this…But I had to take over and deal with it because his brain just could not cope with it (coMND002).

> He gets fixated on things. Like the car's got to be serviced in 10 days, so he's always anxious, I suppose, about some things that are happening (coMND003).

> He gets quite agitated. Sort of like just sometimes simple tasks like, you know, sorting out medications is now it; he gets really quite stressed easily over things like that (coMND006).

> Even though we've been told the diagnosis in the April, he still asked me in the April when he was going to get better... I do not think he ever really, truly understood that side of it (coMND007).

The above quotes bring to life the emotions felt by carers who are experiencing cognitive and behavioural changes.

Sub theme 1.3 - Everyone is different

A strongly held belief of both pwMND and their carers was that each pwMND is different and has an individual journey with the disease. When providing their answers, both pwMND and their carers often prefaced their response with a statement similar to “this is how I feel but it may not be the same for everyone, because everyone with MND is different”. Participants conveyed knowledge that others with MND may have a very different experience to their own, depending on their onset type and rate of progression. There was also an awareness amongst participants that given the variability of MND, people may be at different life stages, and this would also affect their experience.

> …you know, because it does not happen for everybody, and everybody's journey is so different (coMND007).

> …each MND patient has their own journey (pwMND010).

> I've found that with everything with motor neurone because it is just such a different disease with every person (coMND001).

To summarise within the theme, *What I have experience’* and sub-themes, the TDF domains of *Knowledge, Belief about Consequences, Emotion, Environmental context and resources and Social Influences* all played a role.

All participants were aware that dysphagia was associated with MND, and the main symptoms of dysphagia were described as coughing and choking. Further, pwMND also described that eating has become harder and requires more conscious effort. However, some described gaps in their knowledge of dysphagia, specifically about the physiology of swallowing, and the associated risks of aspiration pneumonia when swallowing function is impaired.

Both pwMND and carers described experiencing cognitive and behavioural changes in MND, with reduced empathy, impaired decision making, impaired concentration, impaired reasoning and

increased apathy and agitation reported. This theme also showed that participants had gaps in their knowledge about cognitive and behavioural changes in MND, with some not being aware that these changes could occur in MND until they were interviewed. Additionally, participants described a social influence related to cognitive and behavioural changes, suggesting that this knowledge gap about cognitive and behavioural changes in MND was also represented in the wider community. Additionally, some felt that this was due to a societal stigma related to cognitive decline, meaning that this was less often openly discussed.

Theme 2. What I have experienced related to my care

This theme provides a summary of the experiences families with MND describe about care both within their environment and from HCPs. First, a general overview of care experiences is provided and then experiences of care specific to dysphagia and cognitive and behavioural changes in MND are summarised. Further, subthemes described are 2.1 *What I have experienced in relation to my care for dysphagia*, and 2.2 *What I have experienced in relation to care for cognitive and behavioural changes.* The TDF domains of *Knowledge, Belief about consequences. Social/ professional role and identity, Environmental context and resources, Emotion* and *Social Influences* contribute to this theme.

When speaking about general experiences related to their care, the majority of participants expressed confidence in their professional care team. Most described that they were able to readily access a member of their care team (usually a nurse) as needed. Further, they reported they were kept informed and provided with sufficient information. Many also reported that they felt comfortable asking questions and were given the appropriate amount of time to do so in their clinical appointments. Two participants described the belief that having regularity in their care team created trust and confidence.

Participants frequently discussed various MND services and associations as a reliable source of information within their environment, citing MND services' websites, handouts and association meetings as beneficial to learn about the disease. However, some participants expressed that

impaired mobility and/or distance from an association was an environmental barrier to attending in-person events.

Some participants reported using the internet to research MND. However, one participant described difficulty understanding the information, whilst others expressed concerns about using the internet or social media. These participants cited concerns of misinformation or finding information which causes distress.

> I've tried, but a lot of it is not in lay terms, so it makes it a bit hard because they are studies… (coMND001).

> And sometimes you do not know what's fact, right? (pwMND004).

> I try not to look at Facebook and the groups on Facebook because it's all doom and gloom and it can be quite upsetting when you read that (coMND005).

Some participants reported that at times, their HCPs asked them what they needed, however participants expressed this broad question was difficult to answer as they may not know what they need given the uncharted territory they were facing, or in fact what services or resources were available to them.

> But it's yeah, it's sort of and you know I guess for most people in our position you really do not know what you need until it occurs. So, it's sort of really difficult to know what's coming or and like each person is different. You know, people say, what support do you need? and you go, well, I do not actually know what I need (coMND006).

A contrasting finding was one participant described their belief that the care they received was generalised rather than being specific to their individual situation

> Like most aspects of MND, the focus of professionals is on offering advice (some not always relevant, but apparently standard), techniques or equipment, rather than actually explaining what is going on with my particular degeneration (pwMND012).

Sub theme 2.1 What I have experienced in relation to my care for dysphagia

The majority of participants reported the care they have received from their speech pathologist has helped them to develop knowledge and develop belief in their capabilities to understand and manage dysphagia.

> If I need it, I'm pretty independent. I know pretty much what I need to do with ***. So, I just follow through with what I think. If I've got any problems, I know I can ring them, and we see them once every 12 months, and the (staff) are great. They come up with new ideas, and we discuss things and figure things out (coMND001).

Carers specifically reported that observing a videofluoroscopic study of swallowing (VFSS), in conjunction with education (both verbal and written) was beneficial to build knowledge. They described that observing anatomical structures, residue traces, or aspiration events during swallowing supported their knowledge of dysphagia. Through this, carers described that it helped them to support their pwMND to manage dysphagia outside of the clinic.

> I've been given a lot of print outs by the speech and language people here about how to approach changing your diet, when necessary, what to avoid and what to try (pwMND008).

> I've seen it on diagrams, and I've even seen it when he's had the x-ray type thing done when he's been eating, and they show you the little flap that's supposed to close over and it does not. I do not know all the terminology for it but... And there's this little section here that the food can collect in and that's why you cough and all the things that we take for granted that we can do. So, they've shown me... I have seen it in real-time and I know why it's happening (coMND002).

Whilst the majority of participants described a positive experience to their care for dysphagia, a few participants described feeling not well educated about the pathophysiology and prognosis of

dysphagia in MND. Additionally, two participants described that a lack of information post PEG procedure left them feeling confused.

> I do not think anybody's really explained anything other than what to avoid. And I'm just, I'm, I'm guessing that people would just assume as your muscles become weaker, you realise that that efficiency to swallow becomes, you know, less efficient. Nobody has explained that (pwMND009).

> So, when we went in, got the PEG, they probably did not get very much education or explanation on how to manage the PEG or the wound site. It was sort of here's the food off you go, and we were not really sure what to expect in a bit of weeping. Is that OK? Or you know if it gets infected what that looks like. So that's probably the part that we probably could have done with some more advice or education on that when we left hospital (coMND006).

Sub theme 2.2 What I have experienced in relation to care for cognitive and behavioural changes

When participants were asked to describe their experiences of care specific to cognitive and behavioural changes in MND, some recalled being told that cognitive and behavioural changes may occur for some people with the disease. However, many expressed a lack of communication from their HCPs and reported that this topic either had not previously been discussed with them, or not discussed in detail. Additionally, participants frequently expressed that they felt they were not given opportunities to have general conversations about their disease with their neurologist. Instead, they described their clinical experience mostly involving being asked a series of questions, with little discussion or debrief taking place.

> …no one has ever raised it (pwMND004).

> So, I mean, it's been mentioned, but nobody's sat down and gone right through the whole symptoms of it (coMND001).

> I do not think people have explained anything very much at all (coMND012).

The majority of participants reported that assessment or screening for cognitive and behavioural changes for MND had never been discussed with them, nor conducted. Further, when considering management for cognitive and behavioural changes, all participants interviewed described that they have not been provided with any strategies by their MND care team. Of these, one carer further described her distress when supporting someone who had cognitive and behavioural changes, when not aware that this could occur in MND. The HCP who provided the carer some support in this case was outside of the family's MND care team.

> Now, I was really lucky because I did a mental health care plan with my doctor, and I went to a psychologist, and she was extremely helpful in this area. So, I ended up with a lot of information, which then helped me and ***to understand what was going on (coMND001).

Some participants felt that cognitive and behavioural changes were viewed by their HCPs as not as important as other symptoms of the disease, to either discuss or manage. Additionally, some participants posited that this lack of communication from HCPs may be related to the difficult task of breaking bad news, or for concern that families experiencing MND may not want to receive this information.

> I think medical professionals have to psyche themselves up when dealing with bad news. It is not a pleasant task… (pwMND010).

> That's one thing I was commenting about to someone the other day, that since I've had this disease, those things are certainly changing. But no one actually sat down and said, this is what you can expect could happen. Which is a bit of a pity, I think, but perhaps some people do not want to hear that. I do not know (pwMND002).

To summarise, participants mostly felt confident in their care team and felt they were able to access a HCP when needed. The MND associations were described as a useful resource, however, some participants felt concerned about accessing information via social media or online due to misinformation. Lastly, participants expressed that they wanted to be given specific information and opportunities to ask questions. Participants described that general questions (e.g., what do you need?) were difficult to answer.

In relation to dysphagia care, most participants felt confident in the care they were receiving and reported that, education and visual feedback helped them to better understand swallowing. Some expressed that they were not provided with enough detailed information about dysphagia or about PEG procedures.

Overall, participant's experiences of care for cognitive and behavioural changes in MND were inconsistent, with many reporting that this has not been directly addressed with them. Participants felt that cognitive and behavioural changes seemed less important to their care team, or that their care provider may hold the belief that families with MND do not want to receive this information.

Theme 3. How this affects me

This theme encompasses the personal or relational impact that both dysphagia and cognitive and behavioural changes have on both pwMND and their carers, as reported by the participants. Here, I present the data within the subthemes of 3.1 *The impact of dysphagia in pwMND*, 3.2 *The impact of dysphagia on carers*, 3.3 *The impact of cognitive and behavioural changes on pwMND*, and 3.4 *The impact of cognitive and behavioural changes on carers*. Collectively, the TDF domains that play a role in this theme were: *Belief about consequences, Belief about capabilities, Emotion, Social influences* and *Memory, attention & decision processes.*

Sub theme 3.1 The impact of dysphagia on pwMND

pwMND mostly described the impact of dysphagia as negatively affecting their ability to enjoy foods, as well as being associated with reduced participation in social situations. Participants who were experiencing dysphagia reported a need to be careful when eating and drinking and

described fears of choking and coughing. The most reported food that participants described no longer being able to eat was steak.

> We do not go out as much (pwMND006).

> I just feel sometimes overwhelmed with the quantity of liquid going into my mouth (pwMND009).

In addition, two participants described feeling self-conscious about the use of adaptive equipment in a social environment.

> So just that the food just has to be cut up on the plate and then I can use the robot to select what I want, and it'll bring the food to my mouth. So, I have some independence doing that and we have, we have taken it to a couple of restaurants in the past and I was a little bit anxious about will people be watching me and looking at me. But it's absolutely fine and I have not got any issues about using it outside of the house (pwMND009).

Sub theme 3.2 The impact of dysphagia on carers

Most carers described that their capabilities supported them to feel comfortable managing and supporting their pwMND to continue to eat and drink. Some carers described making adaptations to their own meals to suit the dietary requirements of the pwMND. Further, some carers described that they found decision making challenging, specifically for meal planning. Participants shared the environmental challenges they experienced when trying to find accessible resources, specifically for modified texture recipes or safe food options. Several participants described that this often resulted in the emotion of frustration.

> Some organisation had a recipe book and I thought, Oh, great idea. I clicked on it, and you had to jump through about half a dozen hoops…And I thought, Oh, too hard. I do not want to give them all my personal details and I do not want to give them this, I do not want to give them that just to get this book. Just flipping well give me a book or

> hand something over to me that I can refer to where I do not have to give you my life history or pay for it or pay an exorbitant amount at least with some easy, simple, proven recipes for people who have swallowing difficulties (coMND002).

Many carers reported dysphagia being a barrier to social participation, both related to mealtimes within the home and for social outings. For example, the use of modifying foods (such as the use of thickeners) makes eating out or going to a café difficult.

> So, the whole dynamics has changed. You cannot sit around the kitchen table as easily as you used to and cannot talk as well. So, it's just not very pleasant to be honest that it's got to that point but that's how it is (coMND002).

> …if you go out trying to, you have to thicken your coffee and that sort of thing, which sort of becomes, yeah, yeah so probably has impacted the social interaction quite significantly (coMND006).

Some carers described an awareness that pwMND, who were no longer able to eat and drink by mouth, missed the pleasure that this derives.

> (pwMND) was quite keen to be able to taste things because that's one of the things you really misses is eating and drinking… (coMND012).

> The other day he smells food. You know, homemade. He said put it around my nose (coMND009).

Sub theme 3.3 The impact of cognitive and behavioural changes on pwMND

Participants who reported changes to their cognition and behaviour, described the personal impact this has on their daily living and sense of self. Some reported that they were aware of their altered behaviours but described not being able to control these. Some participants expressed an awareness of how cognitive and behavioural changes impacts on their relationship with their loved ones/carers, and further described feelings of guilt associated with this. Further, participants

experiencing cognitive and behavioural changes also described that their reduced ability to concentrate became a barrier to reading. Some participants expressed that they preferred their carer to read information about MND on their behalf, as they found this task too difficult. Additionally, some participants reported that they had difficulties when driving due to impaired concentration.

> Like the empathy side. I'm definitely, I even know when it's happening, but I've got no control of it. But it's that kind of, it's that insular. I do not really care. Kind of. All right. Whatever. Yeah. In that moment, which is not true *(pwMND turns to his wife)* I do not mean this (pwMND009).

> Definitely has, yeah. Me and Jules built a log cabin together out on the farm, and I do not think we had an argument, but now we argue all the time because I feel I'm right and she's wrong (pwMND001).

> I used to enjoy reading a bit, not that I was a big reader, but I could read quite easily. Whereas now, I really struggle. And not that I cannot read, it's I do not have the concentration to really read much. It's too hard (pwMND003).

> Yeah. It's like a you can get your head around needing to be in a wheelchair quite easily. Lots of people have to use wheelchairs. You can get your head around losing your voice. But this is more difficult because that is part of your personality. Very difficult because that is who you are…Well, I've only just stopped driving recently last December. But if I was trying to navigate through somewhere and there was a lot of traffic, lot of stuff going on, three different conversations in the car. It would, it would really stress me out because all I was focusing on was the driving, and there's all of this background chatter almost that I cannot deal with. If that makes sense (pwMND008).

Sub theme 3.4 The impact of cognitive and behavioural changes on carers

The impact of cognitive and behavioural changes on carers was a significant subtheme within this data. All carers shared individual stories of how cognitive and behavioural changes have impacted them and their families, describing both emotional and financial consequences of these changes. Some carers described their personal experiences of cognitive and behavioural changes before learning this was a consequence of MND. These participants described emotions of confusion, frustration and distress due to the changes in behaviour of their pwMND. In addition, many carers described increased stress associated with the added burden of keeping a pwMND safe from hurting themselves in their environment. Several carers expressed that the cognitive and behavioural changes associated with MND have had a greater negative impact on them than the physical symptoms associated with the disease.

> And to be honest, I find that side of it, the reasoning, the cognitive issues, I find that harder to deal with than the physical disabilities. I can deal with the physical disabilities more than I can deal with the emotional stress that it brings when he's not behaving as he normally would have (coMND02).

> You are taught how to cope because how do you cope with someone who's changed totally? How do you cope with someone who, when you're driving down the road, tells you you're not a good driver, and you need to pass that car, and you're not going fast enough? How do you cope with that when that person told you, *(crying)* when you were younger, you were the best driver? (coMND001).

Further, carers shared personal stories about the financial losses they had sustained, and described ongoing financial impact to their families, after the pwMND deceased.

> He's a naturally generous person and he's always, our whole married life, been very generous with people financially. And I'm having to be a bit of a scrooge now because it's like you cannot keep giving money away (coMND002).

> Well, he, those cognitive impairments have had a huge financial impact. He made some very stupid decisions, very bad financial decisions (coMND007).

> And I think for the carer trying to deal with both the financial loss of perhaps both wages and the mental emotional aspect of losing their, losing parts of their partner and having to take on all the financial responsibilities of that as well as the medical support, caring support. I think there's a huge amount of chaos that is generated and I think denial is one of the easy outs because you can just put it off but actually you cannot, you have to… (coMND010).

One carer described feeling unheard by the MND community when attempting to develop knowledge about the changes she had observed. This resulted in her experiencing increased caregiver burden, which caused an emotional breakdown.

> …every time I kind of brought it up, I was either embarrassed a little bit or was just brushed aside, maybe because I sounded like I knew what I was talking about and was coping, but I was not (coMND001).

Sub theme 3.5 The impact of cognitive & behavioural changes on swallowing

This sub-theme is believed to the be the first time that a link has been established, which shows how cognitive and behavioural changes in MND impact on dysphagia management.

Some carers described the impact of cognitive and behavioural changes in managing dysphagia. Carers reported additional challenges related to ensuring their pwMND was eating safely and following safe eating recommendations. For example, one carer described that her pwMND did not understand dysphagia. Subsequently the pwMND would eat and drink impulsively or would attempt to eat foods which were unsafe. Consequently, the carer described often needing to remove foods.

> …if I find he's choking, I'll just take it off him and go, no, that's not working. So, you're not having that (coMND001).

Further, some carers described beliefs that they needed to regularly remind a pwMND to follow safe eating and drinking strategies or remind them to take smaller portions of food into the mouth.

In summary, the TDF Domains *Belief about consequences, Belief about capabilities, Emotion, Social influences* and *Memory, attention & decision processes* were captured within theme of *'How this affects me'*,

Most participants and carers reported that they felt comfortable managing eating and drinking with dysphagia, and confidence in their abilities to problem solve. However, pwMND reported a reduction in eating pleasure and social participation, and carers described environmental challenges in finding helpful and accessible resources. In contrast however, the impact of cognitive and behavioural changes associated with MND is challenging for both pwMND and carers. Some pwMND reported an awareness of changes to their thinking, which they were not able to control, and described how these changes had negatively impacted on their relationships. In addition, many carers felt confused and distressed, as they did not know the changed behaviour was related to MND. Therefore, these behavioural changes impacted on their relationships. Many also described financial burden associated with cognitive and behavioural changes, which continued to impact on them after the pwMND passed away.

Theme 4. If I had known, it might have been different

This theme relates to the participants' beliefs that if they had received information and/or education about cognitive and behavioural changes associated with MND, their experiences would have been different. The TDF domain of *Beliefs about consequences* featured prominently within this theme. Additionally, *Social influences and Environmental context and resources* were also featured.

The majority of carers, who have lived experience of cognitive and behavioural changes, expressed the view that if they were provided with information about cognitive and behavioural changes early in disease progression, their experience would have been different. Specifically, participants stated that if they had known that cognitive and behavioural changes are associated

with MND, it would have supported their understanding of the disease, and alleviated some of the negative consequences as highlighted in the above theme, How does this affect me? Carers further suggested that having this knowledge would've made caring for a pwMND, who has cognitive and behavioural changes easier, as they would have been able to anticipate changes and understand why these were occurring. Some carers also expressed that having this knowledge would have reassured them about their personal reactions to the pwMND's behaviour changes and brought them comfort that these changes were as a consequence of MND, rather than being a deliberate act from their loved one.

> ...I think it's just really important that you basically that you know what's coming…let's not wait until it's happened. Let's sort of prepare people as best we can…that's one thing that I could change, or that I could, if it's one thing that I think probably needs, look, obviously swallowing is important, otherwise they'll choke. Breathing is important. Otherwise, well, they'll die. But this is just as important. Like, it's one of the big three for this because yeah, as I said, I was not prepared for that part of it (coMND007).

> …but it's like they do not want to worry you beforehand. But if we knew beforehand, I think things can give you a bit of a, you know…it might be actually that this change is starting to happen and you're more prepared then for things. And I think certainly I would not have felt as lost at the start of what's going on with *** and why because it those, those cognition and mental changes really do change a person's personality. And I think initially that is very if you do not if you're not prepared that that actually can go on and I physically have to go and seek that out myself. Why is my husband doing this? (coMND011).

In summary, participants expressed a strong belief that if they had received information that cognitive and behavioural changes may be associated with MND earlier, their experience would've been improved. They believed that learning about cognitive and behavioural changes would have

enabled them to both identify and be prepared for any changes, which may have resulted in feeling more in control and having a better understanding of their situation.

Theme 5. What I do to manage

The data in this theme features the TDF domains of *Knowledge, Social influences, Belief about capabilities, Skills,* and *Emotion.* Here*,* the strategies that pwMND and their carers describe to manage both dysphagia and cognitive and behavioural changes within their daily lives are presented and sit within the following sub-themes: 5.1 *How we manage dysphagia* and 5.2 *How we manage cognitive and behavioural changes.*

Sub theme 5.1 How we manage dysphagia

Most participants felt comfortable in their capabilities to manage dysphagia. Both pwMND and carers explained the skills they had developed to manage changes to swallowing function, with many describing taking smaller mouthfuls and smaller sip sizes. Additionally, food avoidance was frequently described as a strategy to manage dysphagia. Further, a process of trial and error was often described, however, given the progressive decline of swallowing function that is associated with MND, some carers expressed feelings of uncertainty. They reported their experiences of foods that had previously been tolerated by the pwMND may begin to cause coughing and choking as swallowing declines, leading to increased challenges surrounding food choices and decision-making. Additionally, given the variability of dysphagia symptoms experienced MND, the use of straws was described both as a facilitator and as a barrier for safe drinking.

> I avoid a few foods that might set off that choking and coughing (pwMND008).

> There are some things I steer clear of just because they're dangerous to me, like nuts and things, but I can nibble them very slowly, but I cannot just throw a handful in my mouth and eat them (pwMND001).…there was trial and error. You know what I cooked one week he might have been OK with, but a few weeks later he may not have been able to have that sort of a food like (coMND007).

> Probably the most irritating thing was, you know, you'd find something, and you think, yeah, that works well. And then a month later, no, that's no good anymore (coMND012).

Carers frequently described skills they have developed to support their pwMND to continue to eat and drink safely by mouth, such as cutting up food into smaller pieces and adding sauces and gravy to make foods easier to swallow.

> It did change because her eating habits changed. Like if you cut up food, for example, you'd have to cut it up into sizes really really tiny pieces and she'd probably eat as much as a Sparrow because she could not swallow, and she'd cough a lot (coMND008).

> He would have similar to what we were having, but obviously without the taco shells. But everything was just in a bowl because that was easier for him to eat. But yeah, there was definitely a big list of foods that he could not eat anymore (coMND007).

Sub theme 5.2 How we manage cognitive and behavioural changes

Carers described several strategies they enact to manage cognitive and behavioural changes which mostly related to adapting communication and adapting the environment. In relation to adapting communication, some carers described being aware that the pwMND was experiencing rigidity in their thinking and consequently had impaired reasoning. Subsequently, carers described strategies such as 'agreeing to disagree' or ending conversations as a strategy to avoid conflict. Other carers reported altering their communication style to support the pwMND, and here, described conversations which included scaffolded conversations, using repetition and summarising to support the pwMND's understanding. Some carers expressed that they use humour as a strategy to cope with behavioural changes and also described that they attempted not to take the behaviours personally. In addition, carers frequently described attending support groups and having regular conversations with other carers, as a strategy to distract and de-stress from the

carer burden, they were experiencing. Through these conversations they suggested that talking with others in similar experiences resulted in feeling that they were not alone.

In relation to adapting the environment, some carers described the need to hide items around the house which may be unsafe for the pwMND, such as the car keys, ladders, machinery or the details of their bank accounts.

In summary, theme v. W*hat I do to manage* featured the TDF domains of *Knowledge, Social influences, Belief about capabilities, Skills,* and *Emotion*. Overall, participants felt confident in managing dysphagia, and described skills they have developed to manage, such as taking smaller mouthfuls, and cutting up foods into smaller pieces. The main area of difficulty was related to swallowing function decline, which resulted in an increased cognitive load for carers, when they needed to find different foods that were tolerable.

Theme 6. I want care that is relational, individualised, informative and empowering

This theme provides a description of what participants would like their health care to look like more broadly and then specifically as it relates to dysphagia and cognitive and behavioural changes within the following subthemes 6.1 *What care I want for dysphagia*, and 6.2 *What care I want for cognitive and behavioural changes*. This theme features the most TDF domains with *Knowledge, Goals, Emotion, Social influences, Social/professional role and identity, Skills* and *Beliefs about consequences* playing a role in these data.

In general terms, participants expressed a want to be kept informed by their HCPs. Participants also reported that they wanted to learn about all the symptoms of MND. However, some participants also expressed that it was important for HCPs to first determine how much information families experiencing MND would like to know at any given point. They expressed that as everyone with MND is different and has a different experience of the disease, some families may want to receive different levels of information and at different time points. Therefore, they suggested that having regular opportunities to check in and have repeated discussions with their HCPs was an important part of general MND management. Additionally, participants reported that general

resources such as an MND booklet would be helpful to have on hand to refer when they are ready to receive information or to answer questions when they arise.

Sub theme 6.1 What care I want for dysphagia

The majority of participants described that the current care they were receiving from their HCPs in relation to dysphagia was satisfactory and met their needs. One participant expressed that it would be useful to learn first aid skills to support a pwMND in the event of choking. Additionally, some participants reported that they would like to be more informed and receive more education about PEG care post procedure.

Sub theme 6.2 What care I want for cognitive and behavioural changes

All participants expressed that direct education from their HCPs would be their preferred method to learn about cognitive and behavioural changes as associated with MND, stating that this should include general neurophysiology education and prognosis of disease. Most felt that discussions about cognitive and behavioural changes were the responsibility of a neurologist or a general practitioner. Further, many described they desired an opportunity to 'sit and chat' with their neurologist, stating that during this time they wished to receive general education and an overview of MND and be afforded time to ask questions. Taken together, participants described their view that these clinical discussions would help them to better understand the disease and support their understanding of prognosis and progression. However, participants also identified that information exchanges between families and HCPs needed to be conducted in a supportive way, when people were ready to receive information. As such, participants suggested that clinical discussions between HCPs and families with MND need to be individualised, person-centred, and consider personal preferences for learning about MND.

> Rather than health care professionals doing a mental tick list of things they need to impart, the best ones find out how much the patient wants to know, can cope with in one go and to what level of detail they want to understand. I constantly have to battle with senior professionals (the nurses are fine) getting defensive when I ask complex

> questions that they obviously think the patient should not want to delve into (pwMND010).

Additionally, participants expressed that HCPs should ensure that all information is delivered in a way that is comprehensible to an individual's level of understanding.

> Well, for *** and his personality, knowing the way he is, it would just be having that open discussion with him and explain what is actually happening neurologically and what's going on, so he understands (coMND004).

> …depending on the level of impairment, it would be done in a way that they were going to understand it because there are ways that you can ask those, are they understanding it? (coMND007).

The need for an individual approach to discussions about cognitive and behavioural changes was further highlighted by one participant who suggested that learning about cognitive and behavioural changes may lead to feelings of distress and being overwhelmed, and consequently the timing of these discussions is crucial.

> I do think that the information that there might be cognitive change might be a step too far for a lot of people in the early stages (pwMND008).

In addition to verbal education, some participants suggested that written information, in the form of leaflets or handouts, would also be beneficial to support understanding of cognitive and behavioural changes. These participants expressed that having information readily available to refer to within their home environment would be helpful.

To summarise, these data have been presented in the theme of *I want care that is relational, individualised, informative and empowering***.** The TDF domains of *Knowledge, Goals, Emotion, Social influences, Social/professional role and identity, Skills and Beliefs about consequences* contributed to this theme. The majority of participants described a preference for regular open

discussions with their HCPs, with regular opportunities to receive general information that was responsive to their current and individual needs. The majority felt comfortable about the way that their care had been provided for dysphagia. However, for cognitive and behavioural changes in MND, a majority of participants expressed that their experience was different to their preference. Most described their preference for care related to cognitive and behavioural changes involved regular open conversations with their HCPs to receive information. Then, their preference would be to have time to process information via an ongoing discussion with their care team, specifically their neurologist. Further, they expressed that these conversations should be sensitive to their readiness to receive information.

Discussion

In this study, we interviewed pwMND and carers with the aim of elucidating their experiences of dysphagia and cognitive and behavioural changes as a consequence of MND. Additionally, our aim was to understand how families experiencing MND would prefer to receive information, undergo assessment of, and subsequently manage, both dysphagia and cognitive and behavioural changes. We used the TDF framework to explore individual behavioural factors and preferences for assessment and management of both dysphagia and cognitive and behavioural changes. In relation to experience, the following themes were identified: *i. What I have experienced, ii. What I have experienced related to my care, iii. How this affects me, and iv. If I had known, it might have been different.* Additionally, for preferences of care expressed by pwMND and carers, the data revealed two additional themes: *v. What I do to manage, vi. I want care that is relational, individualised, informative and empowering.*

The key findings of this study show that people's experiences of dysphagia management met the needs of the families with MND, whereas this was different to their experiences for managing cognitive and behavioural changes. Families described that their experiences of cognitive and behavioural changes negatively impacted on them and expressed that they if had known more about this symptom of MND, their experiences would've been different. Lastly, participants described their preferences for care, which are in parallel with person-centred care. These findings will now be discussed in detail.

Experiences of dysphagia

Participant's knowledge of dysphagia combined with their functional descriptions mostly align with what is reported in the dysphagia literature (Waito et al., 2017). Functionally, coughing and choking were prominently stated, and participants described the participatory consequences of their swallowing problem related to both reduced food choice and reduced social inclusion. However, despite dysphagia being common in MND, and a serious risk factor for dehydration, malnutrition and aspiration pneumonia, participants mostly described their belief that dysphagia was well managed, and they were confident in their capabilities to cope with the functional changes. Further,

they expressed they were confident in their professional care team, knowing they could reach out if they had a need. Participants also described their care team provided education about swallowing and expressed that observing instrumental assessments helped them to develop knowledge of the functional deficits that were associated with dysphagia. Finally, participants described skills, and a belief about their capability to problem solve dysphagia strategies with trial and error, which they found to be effective. Collectively, the participant's descriptions of the dysphagia care experienced is in keeping with person-centred care models (Hogden et al., 2020) and demonstrates the effectiveness of this type of care to improve patient satisfaction (Australian Commission on Safety and Quality in Health Care, 2023). Namely, patient-centred models of care have been shown to improve patient outcomes when: people feel connected to their HCPs, when they are treated with respect and when they believe they have access to support when the need arises. Further, providing information and education, supports a person's knowledge of the problem and contributes to decision-making (Adams, 2010). Taken together this approach to care promotes the development of self-efficacy, or the belief that one is capable (Paterick et al., 2017). Importantly, in pwMND, self-efficacy has been shown to be positively correlated with satisfaction of life (Galin et al., 2018), even though the person is experiencing a terminal illness. In summary, these findings highlight the importance of a care approach that empowers families with MND to feel capable to manage, whilst also building relationships and partnering with people, to provide individualised support through their MND experience.

Experiences of cognitive and behavioural changes

In contrast to the above, participants described their experiences of cognitive and behavioural changes as vastly different. Despite pwMND reporting knowledge that cognitive changes had personally occurred for them, most of the participants stated that they were not provided with education by their HCPs that cognitive and behavioural changes can be associated with MND. Some participants expressed feelings of guilt related to personality changes and expressed an awareness that these changes are difficult for their loved one to cope with. The feelings of burden felt by pwMND related to cognitive and behavioural changes are less considered in the literature,

with most studies investigating caregiver burden (de Wit et al., 2018). However, it is important to consider the complexities of interpersonal relationships within a family unit when one family member has a terminal disease (McCauley et al., 2021). Furthermore, some carers, reported feeling uncertainty and experiencing distress associated with behavioural changes in the pwMND they cared for. Further, this distress was exacerbated by carers not having knowledge that these changes were because of MND. Additionally, no participants in this study described being given strategies to manage cognitive and behavioural changes from their care team. Most carers described that they independently developed skills to manage, with strategies that included i. changing the home environment, ii. hiding or removing items, iii. drawing on social networks to share experiences and alleviate distress. These findings are not unexpected given the findings described in Chapter 6 & 7, which revealed an inconsistent clinical approach to managing and providing education on cognitive and behavioural changes. Here, HCPs described that cognitive and behavioural screening was seldom conducted, and that educations sessions were not routinely undertaken. One possible explanation for cognitive and behavioural changes in MND not consistently being discussed with families may be due to the TDF domain *Environmental Context and Resources*.

As shown in Chapter 6 & 7, HCPs described environmental barriers specifically related to insufficient time, insufficient funding and insufficient training. Altogether, HCPs suggested clinical management of cognitive and behavioural changes in pwMND were made more difficult because of these factors. Moreover, resource barriers are also reflected in the general health care literature (Wang et al., 2023). Here, it is documented that lack of financial support, staff shortages, inadequate knowledge and inadequate skills are common challenges identified in health care settings. More specifically, resource limitations have been shown to be a barrier to implementing clinical guidelines in health care (Mathieson et al., 2019; Neale et al., 2020; Tan & Black, 2018; Wang et al., 2023). As resource challenges are multifactorial, complex and often outside of the control of the individual HCP, efforts to improve practice for cognitive and behavioural changes in MND which focus on investigating and evaluating realisable ways to overcome these clinical

challenges may in the short term be more beneficial. One such area which may be explored sits within the TDF Domain of *Social Influences* and relates to stigma associated with cognitive decline.

The belief that stigma was associated with cognitive and behavioural change was communicated by participants, with some believing that stigma is present both within society and within their health care team. This belief may be an additional explanation for why clinical conversations about cognitive and behavioural changes in MND may not consistently occur. For example, the findings from both this study and chapter 7 show that it is rare that pwMND are directly asked by their HCPs if they have experienced any changes to their thinking. This evokes some questions, for example: Are pwMND asked directly about their mobility or their swallowing function? Is it more difficult to ask someone about their cognition than their physical functions? How can we support HCPs to better address conversations about cognitive and behavioural changes? How can we educate the broader community about cognitive and behavioural changes in MND to alleviate stigma?

Stigma associated with cognitive impairment is briefly described in the publicly available, Motor Neurone Disease Association (2022) guide ‘Cognitive change and frontotemporal dementia and MND (Pg. 25) and suggests that this may add to the challenge of identifying cognitive and behavioural changes. Whilst stigma is mentioned, the guide stops short on providing solutions for HCPs to work around. Furthermore, ‘Dementia related stigma’, is described frequently in the dementia literature, (Warren, 2023) and suggests that conversations with a person about their cognition may be avoided within society. However, this avoidance can contribute to negative experiences for people who are experiencing cognitive changes, including social isolation, decreased relational closeness, anxiety and depression. Even in mild cognitive decline, people may experience stereotypical and discriminatory behaviours by others, in relation to power inequalities, similar to the experiences of people with a dementia diagnosis (Herrmann et al., 2018). Additionally, mild cognitive decline may change how a person is perceived by others and may lead to social exclusion, due to a belief that this person is no longer themselves or less than they were before (Herrmann et al., 2018; Warren, 2023). Subsequently, a person with cognitive

decline may be devalued. Within this study, some participants believed that HCPs were reluctant to introduce the topic of cognitive and behavioural changes associated with MND, for fear of upsetting or overwhelming people. However, a reluctance to discuss cognitive and behavioural changes with people is not isolated to within the MND context (Warren, 2023). It has been shown to be regularly overlooked in other areas of medical care (Warren, 2023). Within MND research more work is needed to first, evaluate the impact of stigma related to cognitive and behavioural changes in MND. Second, investigations are needed to evaluate strategies aimed at mitigating stigma related to cognitive and behavioural changes in MND to remove this barrier to improve care. However, to add to this already challenging area, cognitive and behavioural changes in MND are not clear cut, as not all pwMND will experience changes to their thinking and behaviour.

As previously stated, approximately half of pwMND will develop cognitive and behavioural changes, however, as these changes are not commonly associated with the disease, some HCPs may not have knowledge of this. Further, some HCPs may believe that it is not appropriate or necessary to provide education on cognitive and behavioural changes to every family, given these changes may not eventuate. Others may hold the view that providing information and education about cognitive and behavioural changes may add to the stress families are already undergoing (Chapter 7). However, despite some HCPs holding the view that pwMND may be overwhelmed by learning about cognitive and behavioural changes in MND, both pwMND and carers in this study expressed that they want to learn about and cognitive and behavioural changes.

Within the theme of *If I had known, it might have been different*, and under the TDF domain *Beliefs about consequences,* carers shared their belief that learning about cognitive and behavioural changes would have helped them to understand the negative behaviours they were observing were because of MND. Additionally, most participants believed that receiving information about cognitive and behavioural changes was an important facilitator to both, develop knowledge of the disease, and to develop their capabilities to be prepared for the challenges that cognitive and behavioural changes may bring. MND guides aimed at supporting HCPs echo this view and suggest that a lack of recognition of cognitive and behavioural changes in MND can increase

stress for carers (MND Association, 2022; Motor Neurone Disease Association, 2022). More broadly, it has been shown that ambiguity within MND care is associated with increased distress, to levels that may, in fact, surpass the distress levels of being told all the information (O'Brien, 2004) (Kutner et al., 1999; MND Association, 2022). Therefore, supporting families through a person-centred care approach may help them to gain a sense of control, and may also help to lessen the emotions of distress, confusion and burden (Oseland et al., 2023) related to cognitive and behavioural changes. Further, this approach may support carer's understanding, and consequently not take challenging behaviours personally, but rather as a consequence of the disease. Taken together, the experiences of both carers and pwMND in this study highlight an important need to incorporate education about cognitive and behavioural changes in MND, with the aim of alleviating the distress that families experience from not knowing why behaviour changes may occur (Pinto et al., 2021). The importance of addressing cognitive and behavioural changes in MND becomes more apparent in the context of family relationship breakdown.

Previous studies have found that pwMND who also have cognitive impairment may have a reduced ability to be empathetic, have a lack of awareness of other's state of mind, and misinterpret subtle signals from their carer (Trucco et al., 2024). Consistent with these reports, participants in our study reported rigidity in their thinking, and described that at times, their immediate needs took precedence over the needs of their carer. Both pwMND and carers also believed as a consequence to this their relationships were negatively impacted. These factors, even when both parties are aware of the existence of cognitive changes, have been shown to lead to both communication and relationship breakdown between a pwMND and their carer (Oseland et al., 2023). This is consistent with the literature describing relational factors in MND when cognitive and behavioural changes are present (Caga et al., 2019). For example, relational closeness is impacted, and reciprocal carer burden can cause psychological stress for both the pwMND and carer (Oseland et al., 2023). Therefore, understanding pwMND and carers preferences for learning about and managing cognitive and behavioural changes appears critical, as this may help both to

better understand the disease, and reduce instances of communication breakdown, having a positive impact on relational interactions.

Families expressed an awareness that at times, they may not know what they need in relation to management strategies or information. Some families expressed certain symptoms of MND they did not see coming. As shown in the theme *How this affects me*, cognitive and behavioural changes were described by some participants as resulting in negative and ongoing financial loss for families. As such, social work services need to be considered in MDT teams to help support families to navigate services related to finances and other legal matters. Both MND clinical guidelines and MND services (Motor Neurone Disease Association, 2022; National Institute for Health and Care Excellence, 2016) suggest that a core MDT consists of a neurologist, specialist nurse, dietician, physiotherapist, occupational therapist, respiratory physiotherapist, speech pathologist and palliative care specialist. In addition, it is suggested that core MND MDT teams have established relationships with additional disciplines, including social work to address social care needs (MND Australia, 2017; National Institute for Health and Care Excellence, 2016).

The importance of a social work service in MND care is highlighted by the experiences of the participants in this study, who described significant financial losses, that they directly associate with not being aware of cognitive and behavioural changes in MND. Whilst it is unclear if the participants in this study had access to social work services, it has been previously shown that families experiencing MND had little awareness that social work may be a service they may benefit from (O'Brien et al., 2012). Given this, it is necessary for HCPs to explain additional services which may help them to navigate the complexities of MND, that may not be directly linked to health care. Additionally, HCPs need to partner with families to undertake supported decision-making conversations, and then advocate for external supports to connect with families.

As described in *sub theme 1.3 Everyone is different,* participants believed that each pwMND experiences the disease differently. Therefore, the ability to predict disease progression is challenging, and the uncertainty of individual MND progression makes providing information more

complex. Consequently, information giving by HCPs needs to be constructed and negotiated with individual families, understanding that some people may want information early, even if this information is inherently bad news, whereas some may find that overwhelming.

The descriptions of what families with MND want for their care in relation to cognitive and behavioural changes in MND align very closely with the findings described in Chapter 5. In Chapter 5, the themes derived from the literature, that describe management of cognitive and behavioural changes, showed that the recommendations were general in nature, however they were situated around assessment (or screening), simplifying communication, conducting early discussions about advanced care planning, providing education and carer support.

Participants in this study described their experiences of care related to dysphagia as mostly meeting the needs of their family. For instance, dysphagia care was described by participants as being based on a trusting relationship, and the provision of education, helped participants to better understand swallowing, and the pwMND's specific swallowing problem. They described developing skills and beliefs about their capabilities, to be able to manage dysphagia within the home environment. Therefore, it can be assumed that adopting this same approach for cognitive and behavioural changes in MND may also be beneficial.

Building trusting relationships with families and seeking to understand preferences for receiving information about cognitive and behavioural changes, may allow for conversations to occur, how and when, it is appropriate for each individual. Further, partnering with families with MND to guide them through supported decision-making processes, will allow them to enact control over their care, which may also help to alleviate some distresses or feelings of uncertainty about what they are experiencing.

Limitations

Some interviews were conducted with both pwMND and carer present to facilitate communication when a pwMND had dysarthria. It is acknowledged that this may have inhibited either participants

ability to speak freely and candidly. Additionally, some pwMND with severe communication difficulties elected to provide their answers to the questions in writing. It is acknowledged that this limited the researcher's ability to ask follow-up questions to clarify any answers.

Additionally, it is acknowledged that the term 'cognitive and behavioural changes' was used in the call out for participants. It is therefore possible that people who responded to this research study did so as they had experienced cognitive and behavioural changes. This may have impacted on the findings.

Conclusion

This is the first study to our knowledge that has considered the impact of cognitive and behavioural changes on dysphagia management. These findings reveal that cognitive and behavioural changes do have negative consequences for pwMND and carers, and impact on dysphagia management. More work is needed to explore this interaction.

Further, participants have strongly voiced that cognitive and behavioural changes in MND are associated with increased burden, for both the pwMND and the carer. This burden relates to increased feelings of guilt and increased stress regarding safety and finances. Additionally, relationship breakdowns were described as a result of misunderstandings caused by behavioural changes. As such, participants believe that education and management of cognitive and behavioural changes in MND is crucial. However, how and when this education is provided needs further investigation.

CHAPTER 9: SUMMARY OF ALL FINDINGS

This chapter provides a summary of the findings from each of the studies included in this program of research.

A first scoping literature review (Chapter 3) identified that there have been no studies that have explicitly investigated the interaction between cognitive and behavioural changes in MND and how these might interact with dysphagia management.

Subsequently, a second literature review (Chapter 5) showed that strategies to guide practice related to managing cognitive and behavioural changes are lacking. Specifically, the existing literature mostly consists of minimal primary data, and management strategies are mostly general in nature, and rely heavily on expert commentaries and evidence that is often derived from outside of the MND context.

In Chapter 6, the findings of an international survey of current practice show that whilst HCPs believe both dysphagia and cognitive and behavioural changes in MND are seen by pwMND as important symptoms to both recognise and manage, there are inconsistencies and irregularities in assessment and management practices for cognitive and behavioural changes. Barriers to screening practices related to environmental factors, specifically with time, skills and staffing identified. Further, HCPs reported not having received any specific training for supporting families with cognitive and behavioural changes in MND. Additionally, HCPs had differing views on whether families with MND would want to learn about cognitive and behavioural changes as associated with MND, with some believing that this may lead to further feelings of being overwhelmed, whilst others felt that families preferred information to feel prepared.

Interviews described in Chapter 7, showed that all HCPs have knowledge that cognitive and behavioural changes can occur in MND, however participants identified gaps in their knowledge related to the neuropathophysiology. They also acknowledged that cognitive and behavioural changes as being associated with MND was often not top of mind. Specifically, HCPs expressed

that each pwMND has a different experience of MND and are often facing severe functional deficits. Further, HCPs described both resource burdens and an uncertainty if families with MND would want to learn about cognitive and behavioural changes as being a barrier to management. Consequently, HCPs manage the more seen symptoms of the disease as a priority.

When considering their current approaches to managing cognitive and behavioural changes in MND, HCPs described providing both verbal and written education. Notably however, all HCPs reported that they had not received training for cognitive and behavioural changes in MND. To this end, they believed that professional development for cognitive and behavioural changes would be beneficial and suggested that training should be inclusive of the pathophysiology of cognitive and behavioural changes, be grounded in MND evidence and draw on the experiences of pwMND and their carers, as well as expert HCPs.

Finally, Chapter 8 describes the experiences of both pwMND and their carers in relation to dysphagia and cognitive and behavioural changes in MND. The findings of this study indicate that cognitive and behavioural changes do impact on dysphagia management. Carers described an increased burden related to cognitive and behavioural changes and managing a pwMND's dysphagia with an increased need to monitor and interject. For example, a pwMND may eat with increased impulsivity and a lack of awareness of the need to eat differently. Additionally, pwMND expressed that apathy impacts on their desire to eat, and carers described a need to prompt their loved one to drink water to maintain hydration.

Notably, families expressed their belief that cognitive and behavioural changes in MND were important to understand to support their coping. Additionally, carers expressed that cognitive and behavioural changes in MND were at times more difficult to manage than the physical symptoms of the disease. However, families described not being given information or education about cognitive and behaviour changes, nor strategies to manage these changes. Further, families did not describe being screened or assessed for cognitive and behavioural changes by any HCP. Most families believed that knowing about cognitive and behavioural changes before they occurred

would have lessened the distress they felt, from not knowing why their pwMND began to behave differently. As such, families believed their preference would be to learn about cognitive and behavioural changes as being associated with MND, early in their MND journey.

Part 3

CHAPTER 10: DISCUSSION

This program of research set out to answer the overarching research question of *How do cognitive and behavioural changes in MND interact with dysphagia management?* As summarised in Chapter 9 and described in Chapters 6, 7 and 8, pwMND, carers and HCPs helped to provide a better understanding of the knowledge about, preferences for and current practices specific to cognitive and behavioural changes and dysphagia. Collectively, these insights showed that the impact of cognitive and behavioural changes in MND is important to understand both in the context of dysphagia management and in the management of MND more broadly. Table 29 shows a summary of the recommendations provided within this chapter, how these align with the TDF framework, and how these may be considered for implementation.

Table 29. Summary of recommendations

Subheading	Recommendation/Solution	TDF Domain	Implementation of recommendation
Cognitive changes in MND and the impact on dysphagia management	Further investigations are needed that explore the interaction between cognitive and behavioural changes in MND and dysphagia	1. Knowledge 2. Environmental context and resources	1. Developing knowledge is necessary to improve care 2. Research is dependent on funding and access to participants
Everyone is different	Clinical conversations need to be tailored to each individual's needs and information provided for all MND characteristics, including cognitive and behavioural changes.	1. Environmental Context and Resources 2. Knowledge 3. Skills 4. Belief about capabilities	1. Opportunities for HCPs to undertake training are needed 2. HCPs to be trained to increase knowledge of cognitive and behavioural changes and communication skills to enact tailored clinical conversations.

Subheading	Recommendation/Solution	TDF Domain	Implementation of recommendation
			3. HCPs develop skills to provide education about cognitive and behavioural changes and adapting their communication style to the individual 4. HCPs feel confident in their capabilities to deliver tailored care
Ethical Considerations	To support a person's autonomy, the HCP needs to understand a pwMND's cognitive abilities	1. Knowledge	1. Develop knowledge of the principles of autonomy in MND care and how cognitive and behavioural changes can impact on decision-making

Subheading	Recommendation/Solution	TDF Domain	Implementation of recommendation
Ethics of Care	Provide care, which encompasses listening to pwMND and their families, to build relationships and trust, and through these relational processes learn of personal preferences	1. Social Influence 2. Environmental Context and Resources	1. Consider the role of relationships in shaping care decisions and the importance of trust between HCPs, pwMND, and families 2. Ensure HCPs have the time, training, and institutional support to engage in relational care.
Reasoning and decision-making	Ask meaningful questions and guide families through decision-making processes Undertake ‘Serious Illness Conversations’,	1. Environmental Context and Resources 2. Skills 3. Memory, Attention, and	1. HCPs to be offered training for various effective communication techniques 2. Implement structured

Subheading	Recommendation/Solution	TDF Domain	Implementation of recommendation
	a “What matters to you?” conversation approach or implement Question Prompt Lists	Decision Processes	conversation frameworks to guide meaningful discussions.

Cognitive changes in MND and the impact on dysphagia management

As described in Chapter 3, the interaction between cognitive and behavioural changes in MND and dysphagia has not been explicitly investigated. However, as shown in Chapter 8, pwMND and carers described their personal experiences of how cognitive and behavioural changes impact on dysphagia management. For example, some pwMND described that apathy meant they no longer had an interest in eating, despite recognising feelings of hunger. Further, carers described that pwMND, who have also had cognitive change, exhibited increased impulsivity when eating and did not appear to understand their swallowing problem. This is consistent with another study that reported pwMND may gorge on food (Trucco et al., 2024), however, is unclear if this impulsivity is related to hyper-orality or impaired understanding and insight. In this instance, carers reported that pwMND continued to eat foods that were of high risk for dysphagia or eat in ways that were unsafe to support safe swallowing, such as taking too much food in a mouthful. Additionally, carers reported that a pwMND did not understand why they were not able to eat all foods they enjoyed prior to being diagnosed with MND. Combined, these examples are consistent with what is reported in the literature describing FTD (Langmore et al., 2007)

As described in Chapter 1, it is valuable to consider the FTD literature in the context of dysphagia in MND, as MND and FTD are understood to occur on a disease continuum (Burrell et al., 2016; Devenney et al., 2015). For example, in FTD, patients are likely to be less aware of their swallowing problem, may eat impulsively and rapidly, and take larger bolus sizes (Langmore et al., 2007). Additionally, it is reported that premature spillage from the oral cavity into the pharynx may occur during mastication and there may be post swallow residue in the pharynx (Waito et al., 2017). As such, these factors contribute to a greater risk of aspiration events and choking for a person with FTD. Importantly, as a link between FTD and MND has been established, these dysphagia characteristics may also present in a pwMND, who also has cognitive and behavioural changes. Consequently, the risk of dysphagia related complications, such as choking or aspiration events, may be further increased in MND, not only due to the motor deficits that impact on swallowing function, but also because of the possible presence of cognitive and behavioural

changes. Taken together, the findings of this program of research raise awareness of the interaction between cognitive and behavioural changes and dysphagia in MND. As such, these findings begin to shed light on some of the negative consequences that may occur for families living with MND, who are managing dysphagia in the presence of co-occurring cognitive and behavioural changes.

Collectively, this program of research shows that changes to current clinical approaches to managing cognitive and behavioural changes in MND are necessary to improve care outcomes and quality of life for both pwMND and their carers. The research undertaken advances our understanding of the interaction between cognitive and behavioural changes and MND management, both from a HCP's perspective and from people with lived experience of MND, as discussed below. Consequently, this research lays the foundation for future investigations about cognitive and behavioural changes in MND.

There is an urgent need to further investigate the interaction between cognitive and behavioural changes in MND and dysphagia, which includes objective measures of both swallowing function and cognitive function, and perspectives and preferences of pwMND and carers with lived experience of MND. Further, when considering cognitive and behavioural changes specifically, this research shines a spotlight on the challenges that are currently present in relation to assessment and management and the complexities which exist within MND care.

Everyone is different

A clear overarching theme that emerged from this program of research is that no two people experience MND in the same way, a view that was shared by pwMND, carers and HCPs. The clinical variations in MND are partly due to the different phenotypes. However, even within the same genetic variant or phenotype, MND disease onset and progression is neither consistent nor predictable (MND Association, 2021b). In addition to varying initial presentations and personal preferences and circumstances, there may also be rapid decline, and symptoms that may occur for some, may not occur for others, or may not occur in a predictable way. Consequently, due to the

vast variability in clinical presentations and individual circumstances, each pwMND will have different needs. The rate of disease progression, the individual functional deficits, as well as individual personal beliefs and wishes for health care all need to be factored into each health care discussion. Additionally, it has consistently been documented that receiving a diagnosis of MND is devastating for both the pwMND and their family (McDermott & Shaw, 2008b; MND Australia, 2024a), and the psychological impact of MND is often associated with feelings of overwhelm, helplessness, anxiety and depression (MND Association, 2023a). Moreover, each individual brings a unique experience related to prior health care experiences, their culture, their values and beliefs. Taken together, this creates complexity for HCPs when providing education about MND to families, as not all people will want to receive education in the same way, or at the same time. Furthermore, it is unclear if some families may prefer to receive no additional information. As a result, and as described in Chapter 7, HCPs may prioritise clinical discussions about the presenting symptoms, rather than risk overwhelming people with information about cognitive and behavioural changes as being associated with MND, as that is not always applicable for everyone. As such, HCPs tailor information that is individualised to meet the preferences of each person.

It is well documented and actively promoted in health care that HCPs, who possess a wealth of expert knowledge, tailor the delivery of health and disease information based on what they determine to be the most relevant for their patient (Smets et al., 2024). As such, a HCP may make choices about what, when and how information is provided (Smets et al., 2024). The overarching aim of tailoring health care discussions is to provide information that meets the individual needs of patients, is relevant to each person's preferences and their capabilities (Smets et al., 2024). In order to do this effectively, HCPs need to first establish trust, and then ask questions designed to learn about personal values and goals for care (Jacobsen et al., 2022). In MND, as the disease is often rapidly progressing, health care discussions additionally need to be dynamic and tailored over time to meet the changing needs of each individual. Notably, within health care discussions, it has been shown that the way HCPs communicate information is of greater importance than the actual information that is being delivered (Xu et al., 2022). How these conversations take place is

directly linked to a patient's satisfaction, understanding and ability to adjust (Luff et al., 2016). Taken together, a clinical approach which does not educate families about all the MND characteristics has ethical implications that require consideration.

Ethical considerations

As outlined in Chapters 6 and 7, some HCPs described that clinical conversations about cognitive and behavioural changes in MND take place if a family broaches this topic, or if the HCP observes changes and other members of the MDT agree with these observations. Some HCPs further expressed their belief that families with MND may find learning about cognitive and behavioural changes overwhelming. Consequently, HCPs described feeling unsure if families wanted to receive early information about cognitive and behavioural changes as being associated with MND. However, information gathering conversations between HCPs and families to determine the families' preferences for receiving information were not described. In Chapter 8, families living with MND additionally reported that education about cognitive and behavioural changes did not occur. This suggests that HCPs may make decisions about which information is important or not important to provide, without knowing the preferences of each individual family. This could be described an as avoidant approach to education about cognitive and behavioural changes in MND.

HCPs expressed feeling uncertain about discussing cognitive and behavioural changes with families, describing their concerns about adding additional burden and overwhelm to families who are already experiencing the grief and burden associated with a terminal illness diagnosis and severe functional impairments. Consequently, whilst HCPs believe the information is important, discussions about cognitive and behavioural changes in MND often do not occur. In contrast, families experiencing MND believed that it is necessary for them to understand that cognitive and behavioural changes may be associated with the disease (Chapter 8). They expressed that having this knowledge would support their understanding of the disease and help them to cope with any changes. This highlights a complexity in health care management of MND, in that HCPs are required to navigate the continuum between paternalistic management approaches on the one hand (i.e., acting in the best interest of each pwMND and their families by not overwhelming them

with information about cognitive and behavioural changes) and principlism management approaches (i.e., enabling self-agency and informed decision making, but at the risk of overwhelming their patient). Consequently, HCPs are acting in, what they believe to be, the best interest of each family. However, it may be suggested that this approach has overlaps with paternalism, which needs to be considered (Beauchamp, 1978). As such, there is tension that exists when considering paternalism in the context of principlism (Hinkley, 2011).

Rooted in the Latin word *pater* (father), paternalism reflects a social hierarchy within patriarchal cultures, where the dominant male makes decisions and choices about their dependent's welfare (Beauchamp, 1978; McCullough, 2016). Acts of paternalism can be related to both actions and omissions of action that come about due to an underlying belief that the action or omission of action will be of benefit to the recipient (Clarke, 2002). In health care, paternalistic acts occur when HCPs make decisions about a person's care, with the belief that their action or omission of an action is in the best interest of the patient. A key aspect of paternalism is that the HCP acts, or chooses not to act, without obtaining explicit and informed consent from the person. For example, if a person is deemed not to have mental capacity, a third-party decision maker or HCP may act to make decisions, which are deemed to be in the best interest of that person on their behalf (Parker, 2004). Further, if a HCP does not present all available treatment options to a patient, but rather presents the treatment they know to be the best available, this would be considered paternalistic in nature.

A common argument which supports a paternalistic approach in health care relates to knowledge. HCPs possess a vast amount of professional knowledge, an understanding of the nature of the condition and knowledge of all available treatment or management options. HCPs use their expertise, combined with expert clinical decision-making processes, to evaluate alternatives to make what they deem to be the best decision for a patient. Additionally, in health care, the patient is often in a vulnerable state and thus is seeking expert advice from the HCP. Consequently, a person may trust the HCP's advice and judgement (Center for Health Ethics, n.d). As such, paternalism in health care posits that i. a HCP may make decisions for, or withhold information

from a patient, if this is deemed in the best interest of the patient, and ii. the HCP knows what is best for the patient. Paternalism is not inherently negative, as paternalistic actions undertaken by HCPs are motivated by a desire to care and protect, but this approach may also limit a person's independence or ability to make an informed choice.

On the other side of this continuum, principlism is a normative ethical framework and common-morality approach to applied ethics, designed to support moral decision-making in bioethics (Beauchamp & DeGrazia, 2004). The four principles underpinning the principlism framework are: i. *Respect for autonomy* (self-determination), ii. *Nonmaleficence* (not causing harm to others), iii. *Beneficence* (acting in a way which prevents harm and benefits the person) and iv. *Justice* (fair, equitable and appropriate treatment of people (Beauchamp & Childress, 1994).

The guiding principle of *Respect for autonomy* broadly involves two conditions i. liberty and ii. agency (Beauchamp & Childress, 1994; Beauchamp & Rauprich, 2016; Bellefleur, 2020). These conditions can be described in relation to a person's right to be free from external constraint and to self-rule. Additionally, autonomy in this context also applies to being free from interference and free from limitations, which prevent individual choice. Autonomy involves the notion that individuals be respected for their individual views and ability to make individual choices. Further, autonomy in health care involves a need for HCPs to support individuals to be appropriately informed, educated, and assisted to enable them to make autonomous decisions (Beauchamp & Rauprich, 2016). Further, the guiding principle of *Nonmaleficence* is grounded in morality that a HCP will not act in a way that will intentionally or negligently cause harm. Similarly, the guiding principle of *Beneficence* is grounded in the morality that HCPs must act in a way that benefits others, as well as preventing or removing possible harms (Beauchamp & Rauprich, 2016; Gillon & Lloyd, 1994). However, when nonmaleficence, beneficence and paternalism are considered in the context of autonomy related to clinical discussions about cognitive and behavioural changes in MND, a degree of complexity becomes apparent.

When considering these principles, there is a conflict that may occur and questions that arise, namely, do health care professionals believe they are acting with beneficence and nonmaleficence by not routinely discussing cognitive and behavioural changes in MND with families*?* And do health care professionals undermine autonomy by acting with beneficence? As such, in the context of this research, it is important to consider whether a pwMND's autonomy is undermined if their HCPs withhold information about cognitive and behavioural changes in MND to avoid causing distress or overwhelm.

Beauchamp and Childress, (2013) discuss their position that beneficence, expressed through paternalistic actions, may be defendable when the action(s) are undertaken for the good of the person who has reduced ability for autonomy. Given that pwMND are often older, have severe functional impairments that impact on both their mobility and communication and may also have cognitive and behavioural changes, autonomy in relation to health care is likely to be impacted (Radakovic et al., 2024; Velaga et al., 2023). This reduction in autonomy in MND is multifactorial and complex. For example, cognitive and behavioural changes associated with MND may manifest as apathy, meaning a pwMND may be less able to initiate and participate in discussions about their own preferences for care. Further, MND negatively impacts on a person's wellbeing, with negative emotions and impaired mental health described (Radakovic et al., 2024). These psychological factors have been shown to further undermine a person's autonomy (Bergamin et al., 2022). Consequently, to support a person's autonomy, the HCP's role is to first understand a person's cognitive abilities, and then work to enable the person to understand and make informed decisions about their care, that aligns with their individual preferences.

It is the role of the HCP to elucidate each person's preferences and undertake clinical discussions that support a person's understanding of MND, whilst at the same time, determining the type and depth of information that they wish to receive. Importantly, the findings described in Chapter 8 demonstrated that not knowing about cognitive and behavioural changes has negative consequences for families, in that relationship breakdowns may occur, and carers may be confused and overwhelmed by a pwMND's changed behaviour. As such, it could also be argued

that not routinely providing information about cognitive and behavioural changes in MND, could be doing harm, in particular to carers. One way for HCPs to navigate the described tensions between paternalism and principlism, is with consideration of an Ethics of Care approach (Gilligan, 1982).

Ethics of Care

Ethics of Care is grounded in both voice and relationships (Gilligan, 1982). Specifically, Ethics of Care considers relational, and context bound approaches towards care and care decision-making (Burton & Dunn, 1996), where every person has a voice, is listened to carefully and heard with respect. Subsequently, the virtues which sit within the Ethics of Care, as described by Tronto (1998) are i. *Attention or Attentiveness* (What care is needed?), ii. *Responsibility* (Who should provide the care?), iii. *Competence* (What are the skills needed and who has these to enact care), and iv. *Responsiveness* (What is the response from the care receiver to the care) (Gilligan, 1982; Tronto, 1998). As such, Ethics of Care principles are context specific, where care is tailored to meet the needs of the individual, rather than implementing a standard approach (Botes, 2000; Gilligan, 1982). This ethical approach supports HCPs to enact care, which encompasses listening to pwMND and their families, to build relationships and trust, and through these relational processes a HCP can learn of personal preferences. Knowledge of these preferences then allows for care to be individualised and inclusive of personal beliefs and wishes. Importantly, Ethics of Care considers the power dynamics that may be present in health care settings when a person who is being cared for is vulnerable (Tronto, 1998). Consequently, an important factor in an Ethics of Care approach is determining how the person being cared for feels about the care they are receiving (Tronto, 1998). However, while this approach addresses some of the need and preferences and overcomes some of the challenges otherwise posed by principlism and paternalism, it also comes with some challenges.

Many barriers to managing cognitive and behavioural changes in MND were identified which undermine an Ethics of Care approach. With regards to iii. *Competence,* cognitive decline has been associated with stigma by both HCP and families with MND (Chapter 7 and Chapter 8), which is a source of conflict for providing care. Additionally, HCPs described resource burdens negatively

impacting on their ability to care for pwMND specifically related to time and funding models. Furthermore, HCPs have not been provided with professional development resources to develop the foundational learning about cognitive and behavioural changes in MND. Taken together, HCPs may lack the knowledge needed to provide information about cognitive and behavioural changes in MND to families, and do not have the resources available to support them to seek out learning opportunities.

When considering the virtue, ii. *Responsibility*, it remains unclear who is responsible for discussing cognitive and behavioural changes in MND with families, meaning that educational conversations about cognitive and behavioural changes occurred inconsistently. Consequently, investigating ways to increase capabilities of the MDT and individual HCPs to provide integrated care is imperative. Within MDTs, HCPs need to work together and combine expertise so that no aspect of MND care, including cognitive and behavioural changes is overlooked. Potentially, an answer to who should be discussing cognitive and behavioural changes in MND is not based on a specific discipline, but rather is based on the development of relationships which are established with mutual trust and shared confidence.

In addition to the identified barriers described above, HCPs are also working with a population of people who are highly variable in their clinical presentation and functional needs, with some experiencing rapid progression, meaning that their functional deficits, and care needs, are also rapidly changing. As such, HCPs need to prioritise and manage many competing demands, within the constraints of health care environments to care for families. Furthermore, because cognitive and behavioural changes in MND may be subtle and masked by other symptoms of the disease, families may not be aware that these changes have occurred, which means these changes may be missed, or mistaken in a negative way. However, as shown in Chapter 8, families experiencing MND, and cognitive and behavioural changes want to know.

Many participants described being unaware that cognitive and behavioural changes could occur in MND and subsequently felt overwhelmed and confused when they or their loved one, began

thinking and behaving differently. Importantly, most also described their belief that their experience would have been different if they had known about cognitive and behavioural changes as being associated with MND, before the changes manifested. Further, participants described the devastating impact that these changes had on their relationships, a pwMND's safety and in some instances, their family's finances. Given this, participants believe it is critically important for families to learn about cognitive and behavioural changes, suggesting that this information should be provided early to support understanding of MND and to facilitate coping. Conversely, as previously described, each pwMND has a difference experience of MND, and consequently each carer also has a different experience of MND. Consequently, some people may prefer not to learn about cognitive and behavioural changes as being associated with MND. In fact, even within families, the preference to learn about cognitive and behavioural changes in MND may be different. Therefore, the complexity for HCPs comes from determining each person's preferences for information in an individualised way that will does not cause additional distress or alarm. Combined, these experiences highlight an important consideration regarding the fundamental rights of families living with MND.

The International Alliance of ALS/MND Association's document describes the Fundamental Rights of People living with ALS/MND and Caregivers (International Alliance of ALS/MND Associations, 2024). This information is underpinned by the United Nations (United Nations, n.d.) and World Health Organisation (World Health Organisation, 2008), and underlying determinants of health, which states that the right to health-related education and information is a fundamental right of all human beings in relation to the right to health. Therefore, families experiencing MND have a fundamental right to receive information that supports them to understand MND and make informed choices in relation to their care (The International Alliance of ALS/MND Associations, 2024). The information provided to families may encompass education and information, as well as providing treatment choices and support in all areas of their care. The provision of access to all information and education about MND enables a pwMND to play an active role in decision-making about their care and support.

Reasoning and decision-making

Given the progressive nature of MND, the functional needs of pwMND change over time, sometimes rapidly. Some symptoms of the disease are subject to windows of opportunity for treatment and as a result, HCPs may present families with prophylactic management choices that require families to make decisions to, or not to proceed before the need arises, such as with PEG insertion for enteral feeding. Consequently, a pwMND may be presented with options and asked to make decisions for their care. However, tensions related to decision-making may occur if a pwMND also has cognitive and behavioural changes.

Cognitive and behavioural changes in MND may present as executive function deficits. These deficits are associated with an increased difficulty in understanding complex information, increased difficulty with decision-making and increased rigidity in thinking (Stojkovic et al., 2016). Further, a pwMND may experience apathy, which may inhibit their ability to engage in decision-making processes (Caga et al., 2018; Caga et al., 2016; Kasper et al., 2015). Therefore, it is critically important for MDTs to establish an understanding of a pwMND's cognitive status, as well as conduct early supported conversations which are in line with the pwMND's cognitive abilities, to guide families through these decision-making processes. Through these supported conversations, HCPs can be confident that a pwMND's choice to proceed, or not to proceed, with treatments or recommendations is based on their own personal circumstance, beliefs and preferences, rather than an inability to initiate or engage in decision making processes for their own care, as a result of cognitive and behavioural changes.

The findings of Chapter 7 demonstrate that HCPs working to support families with MND have not received training that provides essential foundational knowledge about cognitive and behavioural changes in MND and how to address this with families. This highlights an urgent need to develop training resources for all HCPs who are involved in caring for pwMND and carers to learn about cognitive and behavioural changes associated with MND, so HCPs feel supported and have confidence to discuss cognitive and behavioural changes with families. This could include providing direct information to families about cognitive and behavioural changes as well as having

an awareness and providing information about other areas of MND care that may be impacted by cognitive and behavioural changes, such as managing swallowing function.

Speech pathologists apply clinical reasoning processes to assess and reassess swallowing function over disease progression in MND to initially develop appropriate management strategies and then make adaptations as swallowing function changes. Dysphagia management strategies, as described in Chapter 1, require a pwMND to first understand the swallowing problem and then understand and accept or refuse the management strategy. Consequently, managing dysphagia in MND requires a pwMND to cognitively engage with speech pathologists, to understand, learn and then enact dysphagia management strategies within the home. However, without developing an understanding of the pwMND cognitive status, part of the puzzle, which supports clinical decision making, is missing. This further highlights a need for routine discussions about cognitive and behavioural changes to occur as part of ongoing MND management, with HCPs supported to develop meaningful questions to best determine families wishes for their care.

One way to support conversations between HCPs and pwMND/carers is to ask meaningful questions. Within Chapter 8, pwMND and their carers expressed their experience of clinical conversations which did not support them to understand their circumstance or develop an awareness of the resources that might be available to support them. For example, families described difficulty answering the broad question “What do you need?”. Families expressed that they often do not have an awareness of their current or future needs or what services were available. As such, families described their experience of receiving a terminal disease diagnosis as akin to navigating unchartered waters. In this context, it is difficult for families to identify what their needs are or might be in the future. Additionally, each individual within the family unit may have different needs and preferences that arise from adapting to a terminal disease and potentially to new family roles (McCauley et al., 2021). As such, HCPs also need to navigate the complexities that may arise from differing views and differing dynamics within the family unit.

Hence, three possible solutions are provided. The first solution involves training HCPs who support pwMND and carers in how to undertake ‘Serious Illness Conversations’, a relatively new construct developed within the context of palliative care (Jacobsen et al., 2022). For example, The Serious Illness Care Program (Bernacki et al., 2019) developed to support oncology clinicians, provides a training framework aimed at developing the necessary skills to hold supported conversations with patients. The areas targeted in this program aim to help a patient better understand their prognosis, gain patient’s preferences for information, determine patient’s goals, and support patients to express their worries and fears, as well as determine the degree to which they wish family members to be involved in decision-making (Bernacki et al., 2019). This program has been shown to have a positive effect for cancer patients, with improved mood and reduced feelings of anxiety and depression described (Bernacki et al., 2019). These clinical skills are likely to be relevant in the MND context, therefore future research is needed to evaluate this, using codesigned methods.

A second solution may be found by undertaking a “What matters to you?” conversation approach (Barry & Edgman-Levitan, 2012). This approach has been presented as a tool to support an individual’s increased involvement in their own care, aligning with person-centred care approaches (Eklund et al., 2019). Specifically, this framework aims to guide decision-making that aligns with a person’s personal preferences and beliefs (Olsen et al., 2020), and additionally has been used to support people that have cognitive deficits. Further work is needed to determine if this approach to clinical conversations align with how pwMND and carers wish to partake in clinical conversations. The What matters to you? My Goals of care planner (Southern Adelaide Local Health Network, 2024) is shown in Appendix 14.

A third solution to HCPs to determining the individual preferences of pwMND and carers to learn about cognitive and behavioural changes in MND may be via the use of a question prompt list (QPL). QPLs are a set of questions that can be presented to patients with a pre-determined list of questions, which may cover topics such as prognosis, disease symptoms, and treatments and/or management options, as well as addressing the level of family input a pwMND wishes for decision-

making. The use of QPLs in MND may empower pwMND and carers to ask questions and subsequently be more active in conversations about their care. QPLs have been shown to be effective in other populations, such as those with a cancer diagnosis (Brandes et al., 2014; Sato et al., 2022). Further, QPLs may also empower HCPs to instigate serious illness conversations and address sensitive topics, with the knowledge that families have a readiness to receive this information.

In summary, this program of research has shown that there are interactions between cognitive and behavioural changes in MND and dysphagia which need to be considered within a dysphagia management plan, to support both pwMND and their carers to enact safe swallowing strategies in the home environment. For example, both pwMND and carers need to have an awareness of how cognitive and behavioural changes may impact on how a pwMND understands their swallowing problem. Additionally, changes to eating akin to FTD as described above need to be considered. However, more work is needed which investigates this interaction explicitly. The additional findings from this program of research show that there are numerous complexities at play with regards to managing cognitive and behavioural changes in MND, and more work is needed. Specifically, an understanding of the preferences of families experiencing MND are needed in relation to cognitive and behavioural changes. Some possible solutions to these challenges are discussed in the following chapter.

Limitations

Some interviews, as described in Chapter 8, were conducted with both a pwMND and their carer at the same time to facilitate a pwMND's communication when they had dysarthria. It is acknowledged that this may have impacted on each participant's ability to speak freely and candidly.

It is noted that the participants recruited in Chapter 6 and 7 were mostly from an allied health discipline. No neurologists were recruited for either study, so their views are not presented in these

findings. More work is needed to investigate the approaches to assessing for and managing cognitive and behavioural changes, undertaken by neurologists.

It is acknowledged that this program of research has not addressed ways to ascertain the preferences, beliefs and attitudes of pwMND and carers as separate from one another. It is important in MND care, that clinical conversations facilitate ways for pwMND and their carer to communicate with each other and it is likely that families will need to be supported in doing so (McCauley et al., 2021). As described throughout this thesis, cognitive and behavioural changes in MND can impact on a pwMND's communication ability, both due to impaired speech and impaired language due to cognitive changes. In addition, a pwMND may have impaired insight, impaired decision making and may be experiencing apathy. As such, their ability to voice their preferences may be impaired, and thus a pwMND may need to be supported by HCPs to do so. Further adding to this complexity, MND is often associated with dynamic changes within the family unit, meaning people need to assume new roles (Hughes et al., 2005; Lerum et al., 2016; McCauley et al., 2021). These dynamic changes can compound the stress felt by all parties as they navigate managing a terminal illness and the associated functional deficits that occur with MND. To this end, the complex relational interactions that evolve between pwMND and carers when navigating health care in the face of a terminal illness requires further evaluation, to ensure that pwMND and carers are able to jointly participate in clinical conversations and feel supported to express their preferences for care.

CHAPTER 11: FUTURE DIRECTIONS

The process of implementing research knowledge to translate into clinical practice has been shown to take approximately seventeen years from discovery to implementation of knowledge (Morris et al., 2011). Therefore, given that cognitive and behavioural changes were linked to MND in the early 2000s, this knowledge is now only beginning to outgrow its infancy. This possibly explains why training programs for HCPs and clinical management frameworks are currently lacking or very vague.

Within this program of research, the TDF was applied to explore factors that influence behaviour in relation to both dysphagia and cognitive and behavioural changes in MND. By using the TDF, this research provides a theory-driven framework to examine barriers and facilitators to managing cognitive and behavioural changes in MND (Patey et al., 2012).

Specifically, five domains of the TDF were noted to contribute most to the interpretation of the barriers to implementing management of cognitive and behavioural changes in MND by HCPs, namely, i. *Skills*, ii. *Environmental context and resources, iii. Social professional role* and iv. *Belief about consequences* and *v. Knowledge.* To summarise (described in Chapter 6 and 7), *(i.)* HCPs generally do not receive training specific to cognitive and behavioural changes in MND*, (ii.)* have described limited capacity to seek out training, and report resource limitations that impact on their ability to provide care for cognitive and behavioural changes. *(iii.)* Further, HCPs were unsure who should provide information about cognitive and behavioural changes to families and *(iv.)* were unclear of the benefits of addressing cognitive and behavioural changes in MND, believing these conversations may negatively impact on families with MND. *(i.)* Detection of cognitive and behavioural changes mostly relies on family report, observation and/or MDT consensus. Notably, assessment and/or screening for cognitive and behavioural changes in MND is rarely conducted. Additionally, the TDF domain of *(v.) Knowledge* contributed as a barrier to addressing cognitive and behavioural changes in MND. Specifically, many pwMND and carers described that they were not aware of cognitive and behavioural changes as being associated with MND until they began

experiencing these changes either personally or with their loved one (Chapter 8). Further, some of the pwMND and carers who elected to participate in this study, did not have an awareness that cognitive & behavioural changes were associated with MND until the interview was conducted.[12] When asked why this may be, participants expressed their view that there was a persisting misconception within the broader community that cognitive and behavioural functions are spared in MND. Further, some HCPs in Chapter 7 described cognitive and behavioural changes in MND as not being a priority as cognitive function is not thought of being associated with MND, contributing to the inconsistent approaches described. Taken together, this highlights an important need to raise awareness of cognitive and behavioural changes as being associated with MND for both HCPs and the broader community.

Five domains of the TDF were noted to contribute most to the interpretation of the facilitators to implementing management of cognitive and behavioural changes in MND by HCPs. These included i. *Knowledge, ii. Environmental context and resources, iii. Social professional role*, *iv. Belief about consequences* and *v. Goals. (i.)* HCPs know that cognitive and behavioural changes are associated with MND and (*iv*.) believe that managing cognitive and behavioural changes in MND is important. Additionally, *(i. & ii.)* HCPs reported that working in dedicated MDT care teams to support pwMND and carers facilitated their knowledge development. As such, creating MDT teams in MND care that are inclusive of an MND champion who is knowledgeable about cognitive and behavioural changes in MND, provides an opportunity to support and mentor other HCPs within their team, and builds on HCPs' capacity to address and manage this symptom of MND.

The TDF domain of *(v.) Goals* was a facilitator to managing cognitive and behavioural changes in MND and captures important factors to be considered when developing training resources for HCPs in relation to cognitive and behavioural changes in MND. Specifically, HCPs identified that as part of any development of clinical guidelines or professional development programs, practical

[12] (Note: in this instance, education of cognitive and behavioural changes in MND was provided both verbally and with an information booklet)

management strategies and clear communication strategies that are grounded in MND evidence are critical. Additionally, HCPs described that lived experience of pwMND and expert knowledge from other HCPs were important factors to be included in training materials. Finally, HCPs suggested that foundational knowledge about the neuropathophysiology of cognitive and behavioural changes is another important inclusion. Combined, HCPs suggested that these elements would support their knowledge development of cognitive and behavioural changes in MND and provide the necessary foundation that enables them to provide education and support to families with MND about cognitive and behavioural changes. Taken together, these findings provide valuable information which lays the foundation needed to begin the development of professional development resources for cognitive and behavioural changes in MND.

This program of research has highlighted that professional training programs for cognitive and behavioural changes in MND are lacking. As such, there is an urgent need to increase educational opportunities within health care graduate programs about cognitive and behavioural changes in MND. The aim of this inclusion to learning programs would be to arm graduating HCPs with the foundational knowledge needed to build confidence and further enable them to layer new experience dependant knowledge, as they forge their path in a clinical role.

It is hoped that the findings from this program of research, along with the growing body of literature that addresses cognitive and behavioural changes in MND, will help to expediate implementation of knowledge about cognitive and behavioural changes in MND. Specifically, the knowledge generated from this research directly addresses the barriers to management as described by HCPs and describes the current needs of pwMND and carers. Combined, these factors can be considered in a way that leads to improved outcomes for those experiencing MND. However, prior to developing educational programs, more work is needed first to determine families' preferences for learning about cognitive and behavioural changes in MND.

There is a critical need for high-quality research that identifies the preferences of both pwMND, and their carers, related to education about and assessment and management of cognitive and

behavioural changes in MND using co-designed research methods. Undertaking research within co-designed frameworks, which are based on lived experience preferences, will uncover ways of how to work with pwMND and carers to deliver information that it is relevant to their unique experience. Additionally, there is a need to both identify and manage cognitive and behavioural changes in MND and evaluate the benefits of these strategies for families. Within this, it is essential to also develop strategies which allow pwMND who have been identified as having cognitive and behavioural changes to be supported to participate in decision-making processes (Foley & Hynes, 2017). Further, part of this needed research is to determine if early assessment and subsequent identification of cognitive and behavioural changes in MND, with regular repeated assessment to monitor cognitive and behavioural changes, improves care and improves outcomes for families.

A way forward, would be to develop a clinical framework and educational resource that would help families with MND and HCPs to intentionally partner to jointly discuss cognitive and behavioural changes and how they can be best managed in the context of each individual family's preferences, abilities and situation. This is required because as presented in this research, cognitive and behavioural changes are not recognised by the general public to be commonly associated with MND and hence are often not raised by families in clinical conversations. Further, we have shown that HCPs inconsistently discuss cognitive and behavioural changes with families, either due to a lack of MND-specific supporting resources, or because of a perception of potentially overwhelming patients and their families. However, through this research we have also shown that families with MND have a strong preference for learning early on about potential cognitive and behavioural changes. Additionally, families have expressed their desire to be afforded time to sit and chat about MND with a member of their MDT. A possible solution to this could be to create, in some form, a new HCP role, an MND expert care coordinator role.

The vision for an MND expert care coordinator would be to act as the primary point of contact for both HCPs and families throughout the pwMND's entire MND journey. This expert coordinator would both support HCPs in the MDT team, and also serve as a conduit between families and the

greater HCP team. Families would receive regular and ongoing contact with the expert coordinator, who would provide individualised and tailored information, as well as linking families with relevant services, such as social work, and other HCPs as required. Additionally, grief support for the family may be included in this role as part of ongoing care after the pwMND passes away.

Lastly, future investigations, which explore interactions between cognitive and behavioural changes and dysphagia in MND may provide valuable insights into the disease. For example, studies which objectively explore swallowing function in pwMND to determine if there are differences in swallowing function between those with/without cognitive and behavioural changes in pwMND may reveal important insights into disease mechanisms. Additionally, it is of interest in MND research to determine if swallowing function and cognitive and behavioural changes in MND decline at similar rates and if these two symptoms can be correlated. Preliminary research has identified that pwMND experience changes to their eating behaviours (Ahmed, Irish, et al., 2016). Therefore, an area for investigation would be to identify if changes in eating behaviours are an indicator of cognitive and behavioural changes in MND, and whether changes to swallowing function are also identified. Further, studies that summarise and compare and contrast the motor characteristics of dysphagia in MND with the cognitive and behavioural aspects of dysphagia, may be of clinical benefit and create a useful resource for clinicians supporting pwMND.

EPILOGUE

This program of research has touched me deeply. I reflect on times I spent crying with families during their interviews, as they told me they wished their pwMND could live long enough to watch their young children grow. I listened to carers recount their distress from not understanding why their husband was so different to how he was before the disease. Carers, who knew their husband for an entire lifetime, now described that whilst he was still living, he was gone. I also reflect on the changing roles that are necessary within family units in the face of this disease. Women described suddenly having to take over managing the family finances, as their previously competent husband had made decisions that lost the family their life savings. I heard many stories of anxiety and worry about their loved one hurting themselves around the house, and how this meant that they could never be left alone. Men told me that they have now taken on the role of carer, having to feed and bath their wife, because she had not only lost her physical mobility, but also the cognitive capacity to understand how to enact activities of daily living.

I reflect on my admiration for the HCPs who work with families with MND. Their want to protect people from the emotional distress that is associated with losing both their physical functioning and their personality. I imagine the burden that these HCPs must feel at times, working within systems that prohibit for them doing all that they want to, due to time and budget restraints. Our health care system is so heavily funding-centred, which places restrictions on the amount of time that a HCP can spend with a person, and this is often at odds with what people need for their care. Further, within Australia, there is discrimination present for pwMND associated with ageism. Specifically, people who are over 65 years old are not able to access the National Disability Insurance Scheme (NDIS). In contrast, to the current patient-after-patient clinical model, that imposes strict time allocations, many families expressed a need and want to sit and talk to their health care team. To have time to process information. Our current health system often does not permit that to take place.

I recently attended an MND research symposium and sat along some of the greatest minds in MND clinical research. Whilst we have made enormous discoveries in recent times, advancing our knowledge of MND, this group of expert clinical researchers were not confident about discovering a cure within their lifetime. They described MND like cancer, being so complex, with so many disease mechanisms. At this same conference, a man with lived experience provided the most valuable insight in response to this group of experts. He implored them, that until we can find a cure, please focus energy and resources on ways to help pwMND to live their best life, whilst they are still living. This resonates with me so deeply.

So, let us continue to learn from and listen to those who are living with this disease.

Whilst there is no cure, there is care.

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APPENDICES

Appendix 1.
Human Research Ethics Committee Approval Notice, Health care professional (HCP) survey, HCP interviews and person with MND (pwMND) and carer interviews (Chapters 6, 7 & 8)

27 June 2022

Flinders UNIVERSITY

HUMAN RESEARCH ETHICS COMMITTEE

APPROVAL NOTICE

Dear Ms. Rebecca Francis,

The below proposed project has been **approved** on the basis of the information contained in the application and its attachments.

Project No: 4660

Project Title: Investigating cognitive and behavioural change in Motor Neuron Disease (MND)- How is it understood, recognised and managed?

Primary Researcher: Ms. Rebecca Francis

Approval Date: 27/06/2022

Expiry Date: 15/01/2024

Conditions of Approval: None

Please note: *Due to the current COVID-19 situation, researchers are strongly advised to develop a research design that aligns with the University's COVID-19 research protocol involving human studies. Where possible, avoid face-to-face testing and consider rescheduling face-to-face testing or undertaking alternative distance/online data or interview collection means. For further information, please go to https://staff.flinders.edu.au/coronavirus-information/research-updates.*

Please note: *For all research projects wishing to recruit Flinders University students as participants, approval needs to be sought from the Office to the Deputy Vice-Chancellor (Students). To seek approval, please provide a copy of the Ethics approval for the project and a copy of the project application to the Office of the Deputy Vice-Chancellor (Students) via dvcsoffice@dl.flinders.edu.au.*

RESPONSIBILITIES OF RESEARCHERS AND SUPERVISORS

1. Participant Documentation

Please note that it is the responsibility of researchers and supervisors, in the case of student projects, to ensure that:

- all participant documents are checked for spelling, grammatical, numbering and formatting errors. The Committee does not accept any responsibility for the above mentioned errors.
- the Flinders University logo is included on all participant documentation (e.g., letters of Introduction, information Sheets, consent forms, debriefing information and questionnaires – with the exception of purchased research tools) and the current Flinders University letterhead is included in the header of all letters of introduction. The Flinders University international logo/letterhead should be used and documentation should contain international dialling codes for all telephone and fax numbers listed for all research to be conducted overseas.
- the HREC contact details, listed below, are included in the footer of all letters of introduction and information sheets.

  This research project has been approved by Flinders University's Human Research Ethics Committee (Project ID 4660). If you have any complaints or reservations about the ethical conduct of this study, you may contact Flinders University's Research Ethics & Compliance Office via telephone on 08 8201 2543 or by email human.researchethics@flinders.edu.au.

2. Annual Progress / Final Reports

In order to comply with the monitoring requirements of the *National Statement on Ethical Conduct in Human Research 2007 (updated 2018)* an annual progress report must be submitted each year on the anniversary of the approval date for the duration of the ethics approval using the HREC Annual/Final Report Form available online via the ResearchNow Ethics & Biosafety system.

Please note that no data collection can be undertaken after the ethics approval expiry date listed at the top of this notice. If data is

Appendix 2.
Human Research Ethics Committee Approval Notice. Ethnography study. (Chapter 4)

22 February 2021

Flinders UNIVERSITY

HUMAN RESEARCH ETHICS COMMITTEE

APPROVAL NOTICE

Dear Ms. Rebecca Francis,

The below proposed project has been **approved** on the basis of the information contained in the application and its attachments.

Project No:	2850
Project Title:	How do people living with MND understand and manage a swallowing disorder? An ethnographic study.
Primary Researcher:	Ms. Rebecca Francis
Approval Date:	22/02/2021
Expiry Date:	31/03/2023

Please note: *Due to the current COVID-19 situation, researchers are strongly advised to develop a research design that aligns with the University's COVID-19 research protocol involving human studies. Where possible, avoid face-to-face testing and consider rescheduling face-to-face testing or undertaking alternative distance/online data or interview collection means. For further information, please go to https://staff.flinders.edu.au/coronavirus-information/research-updates.*

RESPONSIBILITIES OF RESEARCHERS AND SUPERVISORS

1. Participant Documentation

Please note that it is the responsibility of researchers and supervisors, in the case of student projects, to ensure that:

- all participant documents are checked for spelling, grammatical, numbering and formatting errors. The Committee does not accept any responsibility for the above mentioned errors.
- the Flinders University logo is included on all participant documentation (e.g., letters of Introduction, information Sheets, consent forms, debriefing information and questionnaires – with the exception of purchased research tools) and the current Flinders University letterhead is included in the header of all letters of introduction. The Flinders University international logo/letterhead should be used and documentation should contain international dialling codes for all telephone and fax numbers listed for all research to be conducted overseas.
- the HREC contact details, listed below, are included in the footer of all letters of introduction and information sheets.

This research project has been approved by Flinders University's Human Research Ethics Committee (Project ID 2850). If you have any complaints or reservations about the ethical conduct of this study, you may contact Flinders University's Research Ethics & Compliance Office via telephone on 08 8201 2543 or by email human.researchethics@flinders.edu.au.

2. Annual Progress / Final Reports

In order to comply with the monitoring requirements of the *National Statement on Ethical Conduct in Human Research 2007 (updated 2018)* an annual progress report must be submitted each year on the anniversary of the approval date for the duration of the ethics approval using the HREC Annual/Final Report Form available online via the ResearchNow Ethics & Biosafety system.

Please note that no data collection can be undertaken after the ethics approval expiry date listed at the top of this notice. If data is collected after expiry, it will not be covered in terms of ethics. It is the responsibility of the researcher to ensure that annual progress reports are submitted on time; and that no data is collected after ethics has expired.

If the project is completed *before* ethics approval has expired please ensure a final report is submitted immediately. If ethics approval for your project expires please either submit (1) a final report; or (2) an extension of time request (using the HREC Modification Form).
For student projects, the Low Risk Panel recommends that current ethics approval is maintained until a student's thesis has been submitted, assessed and finalised. This is to protect the student in the event that reviewers recommend that additional data be collected from participants.

Appendix 3.
Scoping review supplementary table I. Full text review articles included for thematic analysis (Chapter 3)

Title	Authors	Theme
A case of late-onset OCD developing PLS and FTD	Bersano et al., (2018)	2
ALS and other motor neuron diseases	Tiryaki et al., (2014)	1,2,3
ALS Three letters that change the people's life. For ever	Oliveira et al., (2009)	1,2
Cerebral amyloid angiopathy and motor neurone disease presenting with a progressive supranuclear palsy-like syndrome	Weeks et al., (2002)	2,3
Clinical variability and female penetrance in X-linked familial FTD/ALS caused by a P506S mutation in UBQLN2	Vengoechea., (2013)	2
Coexistence of amyotrophic lateral sclerosis and argyrophilic grain disease: A non-demented autopsy case showing circumscribed temporal atrophy and involvement of the amygdala	Yokota et al., (2007)	2
Diagnosis of motor neuron disease by neurologists: a study in three countries	Li et al., (1991)	2,3
Diagnosis and management of motor neurone disease	Orrell, (2016)	1,2,3
Dysphagia in amyotrophic lateral sclerosis: impact on Patient Behavior, Diet adaptation, and riluzole management	Onesti et al., (2017)	3
Enteral nutrition in patients with chronic neurological diseases	Caglia et al., (2001)	
ESPEN guideline clinical nutrition in neurology	Burgos et al., (2018)	2,3
Factors associated with communicative participation in amyotrophic lateral sclerosis	Yorkston et al., (2017)	
Factors influencing decision-making in relation to timing of gastrostomy insertion in patients with motor neurone disease	Stavroulakis et al., (2014)	3
Management of sialorrhoea in motor neuron disease: A survey of current UK practice	Hobson et al., (2013)	
Multidisciplinary interventions in motor neuron disease	Williams et al., (2014)	1,2,3
Motor neurone disease: assessment and management	NICE National Clinical Guideline Centre UK (2016)	1,2,3
Nutrition management of amyotrophic lateral sclerosis	Greenwood, (2013)	1,2,3
Nutrition throughout the course of ALS	Golaszewski, (2007)	1,2
Outcome of patients with amyotrophic lateral sclerosis attending in a multidisciplinary care unit	Rivera et al., (2011)	1,2,3
Palliative care in motor neuron disease	Barby et al., (2019)	3
Palliative care triggers in progressive neurological conditions. An evaluation using a multi-centre retrospective case record review and principal component analysis	Hussain et al., (2018)	2
Patters of care for dysphagic patients with degenerative neurological diseases	Sonies (2000)	3
Psychiatric disease in amyotrophic lateral sclerosis.	Maiser et al., (2017)	1,2
Quality improvements in neurology: Amyotrophic lateral sclerosis quality measures.	Miller et al., (2013)	1,2
Taste changes in amyotrophic lateral sclerosis and effects on quality of life.	Tarlarini et al., (2018)	3

Appendix 4.
Scoping review supplementary table II. Coding and categorisation (Chapter 3)

Supplementary Table II. Open coding and categorisations of article excerpt data and the relationship to main theme.			
Raw article exert data	Open code generation	Category	Theme
"…compared 344 patients in MDC to patients in general neurology care (GNC) and found 7.5-month longer survival in the MDC cohort" (Williams et al., 2014).			
"…patients whose symptoms had begun at the bulbar level particularly benefited from multidisciplinary care." (*Rivera et al.,2011).*	Multidisciplinary care improves patient outcomes	Recommendations for care in ALS	Early and regular specialised multidisciplinary management of ALS achieves better outcomes.
"Unfortunately, patients with ALS may not have insight into their personality changes" (*Maiser et al., 2017).* *"Patients are often unaware of any cognitive and behavioral changes"* (*Tiryaki et al., 2014).*	Lack of insight related to cognitive decline		
"Cognitive and behavioral changes can be prevalent and affect the management of patients" (Maiser et al., 2017).	Cognitive impairment impacts on management	Cognitive Impairment	Cognitive decline impacts on management and survival time.
"Shortened survival and a higher rate of non-compliance with recommendations for non-invasive ventilation and feeding tubes are seen in patients with ALS with frontotemporal dysfunction" (Vengoechea et al., 2013).	Cognitive impairment impacts on adherence		

Appendix 5.
Survey question item generation. (Chapter 6)

Management of Cog in MND survey V2

Research Questions

How do practitioners recognise, provide information on, and manage cognitive and behavioural symptoms associated with Motor Neuron Disease (MND) for people and their families living with the disease?

Introduction to the survey

It has been documented that up to 50% of people with Motor Neuron disease will experience cognitive and/or behavioural change (Neary, Snowden, & Mann, 2000; Phukan, Pender, & Hardiman, 2007; Simon & Goldstein, 2019; Watermeyer et al., 2015). A scoping review conducted in 2021 (Francis, Attrill, & Doeltgen, 2021) found that there is limited literature to support clinical decision making on how to manage this symptom of the disease. The purpose of this study is to understand how practitioners and people who support those living with MND recognise the symptom of cognitive &/or behavioural change/s and how they subsequently manage this.

Demographics questions
This section contains general questions about you and your place of work.

1. **Where do you practice?**
   - ☐ NSW
   - ☐ VIC
   - ☐ QLD
   - ☐ TAS
   - ☐ NT
   - ☐ WA
   - ☐ SA
   - ☐ ACT
   - ☐ Other location outside of Australia – free text

2. **What is your gender?**
   - ☐ M
   - ☐ F
   - ☐ Non-binary
   - ☐ Prefer not to say

3. **What is your role?**

   - ☐ Clinical care specialist

   - ☐ Dietician
   - ☐ Gastroenterologist
   - ☐ General Practitioner
   - ☐ Neurologist
   - ☐ Occupational therapist
   - ☐ Palliative care
   - ☐ Physiotherapist
   - ☐ Psychologist/Neuropsychologist
   - ☐ Psychiatrist
   - ☐ Registered nurse
   - ☐ Respiratory Doctor
   - ☐ Social worker
   - ☐ Speech Pathologist
   - ☐ Other (free text)

4. **Which of the follow best describes your clinical setting/workplace?**
   - ☐ Speciality clinic/facility for MND
   - ☐ Private practice with general case load which includes MND
   - ☐ Community Organisation which provides a service for MND
   - ☐ Other (free text)

5. **Is your clinic multidisciplinary (for example nursing and allied health services operating in the same clinic?** Yes / No

The next set of questions ask about your clinical experience.

6. **How long have you been involved in supporting people with MND?**
   - ☐ Less than 1yr
   - ☐ 1-5yrs
   - ☐ 5-10yrs
   - ☐ 10-15yrs
   - ☐ 20+yrs

7. **Approximately how many people living with MND have you treated/worked with?**
   - ☐ 1-5
   - ☐ 5-20
   - ☐ 20-99
   - ☐ 100+

This section asks questions about your views on diagnosing and managing the symptoms of MND and your views on what is important to people living with the disease.

We are interested in how clinicians rate the importance of identifying various symptoms of MND.

8. On a scale of 1 to 5, with 1 being least and 5 being most important, please rate the importance of **diagnosing** the following MND symptoms

(1 unimportant, 2 somewhat unimportant, 3 neither important/unimportant, 4 important, 5 very important)

Respiratory difficulty	1	2	3	4	5
Dysphagia	1	2	3	4	5
Weight loss	1	2	3	4	5
Cognitive and/or behavioural change/s	1	2	3	4	5
Pain	1	2	3	4	5
Sleeping difficulties	1	2	3	4	5
Speech difficulties	1	2	3	4	5
Saliva	1	2	3	4	5
Emotional Lability	1	2	3	4	5
Mobility issues	1	2	3	4	5
Other	1	2	3	4	5

We are interested in how clinicians rate the importance of managing various symptoms in MND?

9. On a scale of 1 to 5, with 1 being least important and 5 being most important please rate the importance of **managing** the following MND symptoms.
   (1 unimportant, 2 somewhat unimportant, 3 neither important/unimportant, 4 important, 5 very important)

Respiratory difficulty	1	2	3	4	5
Dysphagia	1	2	3	4	5
Weight loss	1	2	3	4	5
Cognitive and/or behavioural change/s	1	2	3	4	5
Pain	1	2	3	4	5
Sleeping difficulties	1	2	3	4	5
Speech difficulties	1	2	3	4	5
Saliva	1	2	3	4	5
Emotional Lability	1	2	3	4	5
Mobility issues	1	2	3	4	5
Other	1	2	3	4	5

We are interested in clinicians' views on what people living with MND believe is the most important symptoms to manage in MND.

10. On a scale of 1 with 1 being least important and 5 being most important please rate what is the most importance for people living with MND from the following symptoms.
    (1 unimportant, 2 somewhat unimportant, 3 neither important/unimportant, 4 important, 5 very important)

Respiratory difficulty	1	2	3	4	5
Dysphagia	1	2	3	4	5
Weight loss	1	2	3	4	5
Cognitive and/or behavioural change/s	1	2	3	4	5
Pain / Cramping	1	2	3	4	5
Sleeping difficulties	1	2	3	4	5
Speech difficulties	1	2	3	4	5
Secretion/Saliva	1	2	3	4	5

Emotional Lability	1	2	3	4	5
Mobility issues	1	2	3	4	5
Fatigue	1	2	3	4	5
Other	1	2	3	4	5

The next section asks questions specifically related to cognitive and behavioural change/s that are known to be associated with Motor Neuron Disease. We are interested in understanding how this is identified and managed in people living with MND.

11. **Have you ever observed cognitive &/or behavioural change/s in people with MND?** Y / N

12. **What processes/procedures do you have to recognise or detect cognitive &/or behavioural change/s in a person with MND**? Free text

13. How do you detect cognitive&/or behavioural changes in people with MND who have a communication impairment (anarthria/dysarthria)? Free text

14. **How often do you discuss cognitive &/or behavioural change/s as a symptom of MND with people living with the disease their families**? Always Sometimes Rarely Never

15. **How comfortable are you discussing cognitive &/or behavioural change/s with families living with MND** V U, U, Neither U or C, C, VC

16. **How often are you asked for information about cognitive &/or behavioural change/s associated with MND by plwMND?** Always Sometimes Rarely Never

17. **How often are you asked for information about cognitive &/or behavioural change/s associated with MND by families?** Always Sometimes Rarely Never

18. **In your view, do plwMND want to know about cognitive &/or behavioural change/s?**
    a. If not, why not?
    b. If yes, why yes?

19. **In your view, do families of plwMND want to know about cognitive &/or behavioural change/s?**
    a. If not, why not?
    b. If yes, why yes?

20. **In your view, who is responsible for discussing cognitive &/or behavioural change/s associated with MND to people with the disease and their and families? (Check all that apply)**

- ☐ Clinical care specialist
- ☐ Dietician
- ☐ Gastroenterologist
- ☐ General Practitioner
- ☐ Neurologist
- ☐ Occupational therapist
- ☐ Palliative care
- ☐ Physiotherapist
- ☐ Psychologist/Neuropsychologist
- ☐ Psychiatrist
- ☐ Registered nurse
- ☐ Respiratory Doctor
- ☐ Social worker
- ☐ Speech Pathologist
- ☐ Other (free text)

21. **Please briefly describe what and how you explain cognitive &/or behavioural change/s to people with MND and their family/carers?**
    Text

22. **How often do you or your service provide education, resources, or further information regarding cognitive &/or behavioural change/s in MND to plwMND and families?**

    Always Sometimes Rarely Never

23. **What information do you provide? (check all that apply)**
    - ☐ verbal education
    - ☐ hand-out prepared by your service
    - ☐ hand-out prepared by an MND organisation
    - ☐ website links
    - ☐ Other (free text)

24. **How often do you or your service recommend/refer families with MND to community-based groups for support with cognitive &/or behavioural change/s?**

    Always Sometimes Rarely Never

The next section relates to education and information for clinicians about cognitive &/or behavioural change/s in MND

25. **Have you received specific training on how to support someone with cognitive &/or behavioural change/s in any neurodegenerative condition?** Y/N

    a. If yes, can you describe this training?

26. **Have you received specific training on how to support someone with cognitive &/or behavioural change/s specifically related to MND?** Y/N

    a. If yes, can you describe this training?

27. **How do you access information about cognitive and behavioural change in MND?** Text

We are interested to know what resources are available to clinicians to learn about cognitive and behavioural change in MND. Please provide links or upload information for yourself or your practice about cognitive and behavioural change in MND. For example, clinical guidelines and/or pathways, online resources, MND association handouts. Any information provided will be treated confidentially and without publication of materials. It will be solely used for research purposes to map the types of information that is available.

28. Drop box /upload box **INCLUDE LINK**

The next section relates to cognitive and behavioural screening practices in MND. We are interested in MND specifically but also how these practices compare to other neurodegenerative conditions.

29. As part of routine practice, does your service conduct cognitive &/or behavioural screening in the initial contact with a person with confirmed or probable MND? Always Sometimes Rarely Never

    a. If yes, which screening tool is used? List

30. As part of routine practice, how often does your service conduct cognitive &/or behavioural screening in the initial contact with a person diagnosed with other neurodegenerative conditions? Always Sometimes Rarely Never, Other (free text)

    a. If yes, which screening tool is used? List

31. As part of routine practice, does your service conduct follow up screening review at regular intervals for cognitive and behavioural change in MND? Yes/No

32. Does screening review for cognitive &/or behavioural change/s take place at regular intervals? Yes/No
    a. If yes, how often do you conduct a screening review? 3mths, 6mths, 12mths, Every apt, Other (free text)

33. As part of routine practice, how often does your facility conduct follow up screening review at regular intervals for cognitive &/or behavioural change for other neurodegenerative diseases? Always Sometimes Rarely Never, Other (free text)

    a. If yes, how often do you conduct a screen? 3mths, 6mths, 12mths, Every apt, Other (free text)

The next section asks you questions related to cognitive screening in MND only

34. In your view, how important is it to formally screen for cognitive &/or behavioural changes in plwMND? Likert scale (1 not important, 5 Very important)
    a. Please explain your rating response (why have you given the answer you have given)? Text

35. In your view, are there benefits for screening for cognitive &/or behavioural changes in plwMND? Y /N
    a. If yes, why / If no, why not Text

36. In your view what, if any, are facilitators for regular screening of cognitive &/or behaviour changes in MND in your role? Text

37. In your view what, if any, are barriers for regular screening of cognitive &/or behaviour changes in MND in your role? Text
    a. Please identify any solutions to the barriers you have identified. Text

This brings us to the end of the survey. Thank you for taking part.

38. Is there anything else you would like to comment on? Text

We are offering participants the option to be involved in a follow-up semi-structured interview to expand on the answers you have provided. This session will take approximately 20 minutes of your time and you will be offered a $40 online gift card for your time.

39. Please provide your email address if you consent to be contacted Text

Suggestions for reflection questions from study:

How is cognitive and behavioural screening currently addressed in your practice, and how might it be improved?

Test of survey to be completed with:

MND SA staff member, Speech Pathology staff (Lara Ferris, Mystika) Nutrition, Physio, David Shulz? Peter Allcroft? Mark Slee Flinders.

References

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Phukan, J., Pender, N. P., & Hardiman, O. (2007). Cognitive impairment in amyotrophic lateral sclerosis. *The Lancet Neurology, 6*(11), 994-1003. doi:https://doi.org/10.1016/S1474-4422(07)70265-X

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Appendix 6.

Survey distribution list. (Chapter 6)

Appendix 7.

Email script to HCPS via publicly available email addresses. (Chapter 6)

Dear Practitioner,

I am writing with regards to a brief survey that I am conducting as part of my PhD studies at Flinders University, South Australia. My name is Rebecca Francis. I am Speech Pathologist and as part of my PhD project, I am conducting research investigating how the symptom of cognitive and behavioural change in Motor Neurone Disease is managed by practitioners who support families living with the disease.

You are invited to participate in this research project by completing a short questionnaire. A full information sheet about the research project is attached. It is anticipated that completion of the questionnaire will take approximately thirty minutes. Any information you provide will be treated strictly confidential and neither you or any of your responses will be identifiable in the resulting thesis or publication.

You are, of course, entirely free to discontinue your participation in the questionnaire at any time or to decline to answer any of the questions. You will be required to provide your consent to take part in the research project, which will be done by completing the first question on the questionnaire.

You can complete the questionnaire by following this link xxxxx

Any enquiries you may have concerning this project should be directed to myself by telephone on (08) 8201 2811 or email rebecca.francis@flinders.edu.au

Thank you for your consideration.

Rebecca

This doctoral research is supervised by Associate Professor Sebastian Doeltgen (Flinders University) (sebastian.doeltgen@flinders.edu.au) and Associate Professor Stacie Attrill (University of Adelaide) (stacie.attrill@adelaide.edu.au)

Appendix 8.

Interview Guide. Health care professional interviews. (Chapter 7)

How is cognitive and behavioural changed managed in MND?
Part A - Practitioner survey

Topic Guide for Semi structured interviews

The practitioner semi structured interviews which are within Part A of this research project will be mostly practitioner lead.

These interviews will be held at a time that is convenient to the practitioner as determined via Doodle Polling.

The email script to Practitioners is as follows:

Dear Practitioner,

Thank you again for completing my survey and providing your details for the follow up interview.

I am hoping to meet with you week commencing the 7th or 14th Nov. I have created a Doodle Poll with a link here: https://doodle.com/meeting/participate/id/bmOZyX9e

If you would ever so kindly indicate on this poll when you would be available, and I will then email you a Zoom/Microsoft Teams meeting link.

If none of the times/days suggested are suitable I am more than happy to schedule a time that is more convenient to you. Please advise via return email when a suitable day/time would be.

Should you have any questions, please contact me via return email or on +61 82012811

Warm regards
Rebecca Francis

Interview Process

The interviews will be held online via Zoom or Microsoft Teams (or another platform that is preferred by the practitioner). The semi structured interviews will be scheduled for one hour. At the beginning of the session, the CI will ask the practitioner for consent to record the session to allow for data review and transcription. The practitioner will be advised at the beginning of the interview session that they can end the interview at any stage, or if they wish they can continue to give answers past the allocated one-hour time frame. The CI will bring the interview to a close if it goes over 1hr and 30 mins.

The lead investigator will ask guiding questions which will be centred around the data collected from the survey.

The guiding questions for the semi structured interviews are as follows:

1. CI - *"From my data I can see that 70 % of practitioners conduct a screen for cognitive and behavioural changes at least some of the time. Can you describe what might happens after this screen has been conducted in your clinical setting?"*

RQ – What do you do to manage cog and behavioural in your routine practice?

RQ – what are the care pathways for managing cog and behavioural change in MND

2. *CI - "From my data I can see that approx. 24% of practitioners felt "somewhat uncomfortable" and 33% felt "somewhat comfortable" talking about cognition with families. From your perspective why might this be and what do you think would improve this for practitioners?*
    a. *Prompt – in your experience what makes discussing this more uncomfortable? More comfortable?*
    b. *Prompt – in your experience how do they receive this information? How do you find they generally react to learning about this symptom?*
    c. *If they have been screened to have change, how do you find they react to the news they have experienced cognitive/behavioural change?*
    d. *Do you think stigma exists in this space?*

    RQ – What are practitioners ?

3. *CI – "From my data I can see that approx. 45% of respondents provided education for cognitive and behavioural change associated with MND, and much of this education (over 34%) was provided verbally only. Can you tell me about the provision of education related to cognitive and behavioural change in MND specific to your setting?*
    a. *Prompt - In what medium is this sharing of information done?*

4. CI – *"Many respondents in the survey described a multi-disciplinary approach to managing cognitive/behavioural deficits in MND. What does that mean/look like in your setting?"*
    a. *Prompt –disciplines? / barriers? / processes?*

5. *CI - Only approx. half the respondents in the survey report having received training for supporting someone with cognitive/behavioural changes related to any neurodegenerative disease. Have you received training? If Yes - Can you describe this? If No – Can you suggest why this might be?*
    a. *What do you think needs to be in the training?*

6. Upload of Clinical pathway tools – *We are interested in how practice is similar and different across locations, settings and disciplines. Anything additional that you can provide re resources, websites that we might be able to collect as an opportunity to collate the breadth of literature.*

7. Is there anything else you would like to add personal or professional?

Address for gift card to be posted out

Thanks for time. Results of this research can be sent to you if you wish to receive.

Appendix 9.

Recruitment flyer pwMND and carer interviews. (Chapter 8)

Appendix 10.

HCP Participant information and consent form. (Chapter 7)

How do practitioners recognise, provide information on, and manage cognitive and behavioural symptoms associated with Motor Neuron Disease (MND) for patients and families living with the disease?

Welcome.

Thank you for recently completing the online survey and providing your contact information. I would now like to invite you to participate in a discussion, in the form of a semi-structured interview to expand on the information collected in the survey. This semi-structured interview aims to further explore the clinical conversations and information sharing between practitioners and families living with Motor Neuron Disease about cognitive & behavioural decline.

This study forms part of a Rebecca Francis' PhD project that is investigating the impact of cognitive & behavioural decline associated with Motor Neuron Disease on swallowing.

To participate you must be a practitioner who works with an MND population.

Before you decide to take part, it is important for you to understand why the research is being conducted and what it will involve. Please take your time to read the participant information sheet page carefully. Participation is voluntary. Please contact us if anything is unclear or if you need more information on the contact details provided.

Research Team Chief Investigator
Ms. Rebecca Francis
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ph. 08 82012811

Research Supervisors
- Associate Professor Sebastian Doeltgen sebastian.doeltgen@finders.edu.au
- Associate Professor Stacie Attrill stacie.attrill@adelaide.edu.au

What is this research about?
Some people living with MND may experience cognitive and behavioural change as a symptom of the disease. Currently little is known about the clinical pathways that are in place to manage this symptom of the disease. Therefore, the purpose of this study is to understand how practitioners, who work with families living with MND, recognise and asses for the symptom of cognitive and behavioural change/s, how they provide education on this symptom and how they subsequently provide support and management.

What benefit will I gain from being involved in this study?
The sharing of your experiences will help us learn more about how information related to cognitive and behavioural change in MND is provided to families living with the disease. The findings from this study may help to improve clinical care for families impacted by MND.

Recognition of Contribution
If you would like to participate, in recognition of your contribution and participation time, you will be provided with a $40.00 Coles Group & Myer gift card. A physical gift card will be mailed to you on completion of the interview. Subsequently, you will be required to provide a postal address to receive this.

Appendix 11.

People with MND and carer interview guide. (Chapter 8)

Investigating cognitive and behavioural change in MND. How is it understood, recognised and managed?

Part B interview questions

Approved ethics proposal #4660

RQ - How do pwMND & carers experience the symptoms of dysphagia and cognitive and behavioural decline in MND, and how would they like these symptoms to be managed?

(Part A of this projected consisted of a survey and follow up interviews with practitioners working with MND. This part of the study has now been conducted and data analysis has begun)

Part B consists of semi structured interviews to be conducted with people with MND, family members of someone with MND and/or primary carers of someone with MND. Participants will be offered the choice of being interviewed together or separately. Carers will also be offered the opportunity to have a follow up chat by themselves if they wish.

The World Health Organisation International Classification of Functioning (WHO, 2020) will be used as a conceptual framework to explore experiences and attitudes.

The following outlines the general interview guide with proposed interview questions as follows. Note that these questions will be asked as part of the flow of the interview and may be asked in a different order. The wording of questions will be adapted to the interviewee(s)' situation, including reference to partner/spouse/name and information will be provided where concepts or questions are unclear.

This first set of questions are about you.

Questions for PW MND	Questions for CARER OF PWMND
Where do you live?	**Where do you live?**
What is your age?	**What is your age?**
M/F	**M/F**
How long ago did you receive your diagnosis?	**How long ago did your *loved/partner/spouse (as appropriate)* one receive their diagnosis?**
Do you know the type of MND you been diagnosed with?	**Do you know the type of MND your loved/partner... one has been diagnosed with?**

I am now going to ask you some questions about your swallowing:

Nature of the behaviour	
Tell me about your swallowing. How has it changed since your diagnosis? **Participant swallowing safety questions.** Have you had any speech pathology or other practitioner support for this? a. If NO – recommend a follow up with their medical support (GP,	**Tell me about how your loved one's swallowing. How has it changed since their diagnosis?**

neurologist and/or Speech Pathologist) Have you had your swallowing assessed? (What was the outcome?) Have you ever coughed or choked while eating? b. If YES – recommend a follow up with their medical support (GP, neurologist and/or Speech Pathologist)	
Tell me about how you have changed things to help your swallowing? What do you normally do? Prompt – changes to food, mealtimes, going out etc.	**Tell me about how you have changed things to help your loved one's swallowing?** What do you normally do? Prompt – changes to food, mealtimes, going out etc.
Social/professional role and identity	
Who supports you with your swallowing? Prompt – which professionals, within the home	**Who supports your loved one with this?** Prompt – which professionals, within the home
Are there any challenges around this?	**Are there any challenges around this?**
Do you feel like your swallowing problem has been adequately explained to you?	**Do you feel like your loved one's swallowing problem has been adequately explained to you?**
Social Influence	
Do you feel like your carer/partner/spouse understands your swallowing problem. Has this been adequately explained to them?	**Do you feel like the person you care for understands their swallowing problem? Has this been adequately explained to them?**
What recommendations have you been given?	**What recommendations have you been given?**
To achieve these recommendations, what needs to be done differently? (e.g., do others need to do something? Something new is needed?)	To achieve these recommendations, what needs to be done differently? (e.g., do others need to do something? Something new is needed?)
Skills	
How easy or difficult do you find it to follow the swallowing recommendations?	**How easy or difficult do you find it to follow the swallowing recommendations?**
What could be done to make it easier?	**What could be done to make it easier?**

We know that for some pwMND, there can be changes to cognition and behaviour. Cognition refers to the thought processes or thinking and include things like reasoning, awareness, perception or remembering. So this can means some people with MND may need extra support with things like problem solving or understanding new information. The next set of questions are related to this:

How familiar are you about the symptom of cognitive and behavioural change in some people with MND?	**How familiar are you about the symptom of cognitive and behavioural change in some people with MND?**
Knowledge	
How did you learn about this?	**How did you learn about this?**
Would you want to find out if this was the case for you? Omit next question if answer No here	**Would you want to find out if this was the case for your loved one?** Omit next question if answer No here
How would you want to learn about this? (If a prompt is needed – for example from your neurologist, from reading about MND on the internet, from the MND association etc)	**How would you want to learn about this?** (If a prompt is needed – for example from your neurologist, from reading about MND on the internet, from the MND association etc)
What do you think are the benefits of this for you? For your family?	What do you think are the benefits of this for you? For your loved one?
Social Influence / Environment / Social professional role and identity	
Who do you think should discuss this symptom of the disease with you? (Prompt –GP, Neuro etc)	**Who do you think should discuss this symptom of the disease with you?** (Prompt – GP, Neuro etc)
In your opinion, when do you think would be the best time to learn about this symptom of the disease? (Prompt – when first diagnosed, or later, e.g. when first symptoms appear?)	**In your opinion, when do you think would be the best time to learn about this symptom of the disease?** (Prompt – when 1st dx, later?)
In your opinion, how should a clinician discuss this with you? What would be your preference?	**In your opinion, how should a clinician discuss this with you? What would be your preference?**
What do you think would be helpful to learn about this symptom of the disease? (Prompt – handouts, conversation, websites, videos, other people's stories etc)	**What do you think would be helpful to learn about this symptom of the disease?** (Prompt – handouts, chat, websites, videos, other people's stories etc)
How often would you like to discuss this with your practitioner?	**How often would you like to discuss this with your practitioner?**
Knowledge	
Have you accessed any resources on this information yourself? **If yes – can you describe this**	**Have you accessed any resources on this information yourself?** **If yes – can you describe this**

The final set of questions is asking you about how to manage cognitive and behavioural change.

2. Have you experienced practitioners explaining information to you in a way that wasn't clear?
3. What could they have done differently to make it clearer?
4. Do you feel you were given enough time to clarify the information? (e.g., ask questions etc)
5. What recommendations, if any, have you been given to manage cognitive & behaviour change?
6. How easy or difficult do you find it to follow these recommendations?

7. What could be done differently? What would be helpful for you to manage cognition and behaviour changes?
8. What do you think would be important for clinicians to do if you/the person you care for was found to have cognitive and behavioural change?
9. Has your practitioner discussed screening for cognitive and behavioural change with you/your loved one?

Appendix 12.

Participant information and consent form, people with MND (Chapter 8)

INFORMATION SHEET AND CONSENT FORM – PARTICIPANT

How do people living with Motor Neurone Disease and their primary carers learn about, understand and manage cognitive and behavioural change?

Chief Investigator
Ms. Rebecca Francis
College of Nursing & Health Science
Flinders University
Tel: 08 82012811
E: Rebecca.francis@flinders.edu.au

Research Supervisor
Associate Professor Sebastian Doeltgen
College of Nursing & Health Science
Flinders University
Tel: 08 7221 8817

Research Supervisor
Associate Professor Stacie Attrill
External Organisation
Tel: 08 83133518

Description of the study

Chief Investigator Rebecca Francis is a PhD candidate at Flinders University, and this study will form part of her doctoral research program. We are inviting you to consider participating in a research project, which will explore your experiences of learning about, receiving education on, and if applicable, managing cognitive and behavioural change that can be a symptom of Motor Neuron Disease.

This Participant Information Sheet/Consent Form tells you about the study. It explains how the research will be done. Knowing what is involved will help you decide if you want to take part in the research.

Please read this information carefully. Ask questions about anything that you don't understand or want to know more about. Before deciding whether or not to take part, you might want to talk about it with a relative, friend or your local doctor.

Participation in this research is voluntary. If you don't wish to take part, you don't have to. You, or the person you care for, medical care, relationship with the university and/or the services which are received will not be affected by your decision.

If you decide you want to take part in the research project, you will be asked to sign the consent section. By signing it you are telling us that you:

- Understand what you have read
- Consent to take part in the research project
- Consent to participating in the screening assessment that is described

You will be given a copy of this Participant Information and Consent Form to keep.

This project is supported by Flinders University, College of Nursing and Health Science.

Purpose of the study

Some people living with Motor Neuron Disease may experience changes in their cognition, such as changes in thinking and problem-solving skills and/or their behaviour. Currently there is no information that tells us how people living with Motor Neurone Disease and their carers learn about this symptom of the disease, what education they receive, and how they are supported to manage these symptoms. It is also unclear how these symptoms may impact on how plwMND and their families manage swallowing difficulties. This research will explore your experiences related these symptoms.

We will do this by conducting semi-structured interviews with people who have MND and/or with people who care for someone who has MND. We anticipate that this interview will take approximately thirty minutes. Please note you can decide at any point, including during the interview to withdraw from the research or discontinue the interview.

During the interview, the lead researcher will ask you some general questions about your experiences with MND, questions related to the symptom of cognitive and behavioural change and questions about your swallowing function and eating and drinking.

Benefits of the study

By sharing your experiences, you will help us understand how plwMND and their carers learn about symptoms of the disease, what educational resources they receive and how they like to receive this information. In addition, this research will help us understand how swallowing is managed every day, and if cognitive changes impact on swallowing management for both the plwMND and their primary carer.

By collecting this information, we will be able to improve health services for plwMND and their families. It will also provide education to health practitioners to assist them in making decisions and to help them to improve the care they provide.

We cannot guarantee or promise that there are any direct benefits to you from this research, however possible benefits may include more help for people to manage and make decisions about their cognitive and swallowing management in the future. There is little information about how people manage their swallowing during the course of MND, and this study will provide important information about this.

Recognition of Contribution / Time / Travel costs

If you would like to participate, in recognition of your contribution and participation time, you will be provided with a $40.00 Coles Myer gift card. This gift card will be posted to you on completion of the interview.

Participant involvement and potential risks

A member of the research team will give you information about the study. If you choose to participate in the study, you will need to sign a consent form. If you choose not to participate, you will not be contacted again by this research team about this study.

If you consent to participate in this study, you will be invited to attend an online interview via Microsoft Teams or Zoom or on the telephone. Alternatively, if it is your preference, a one-on-one face to face interview can be arranged at a location that is convenient to you. All interviews will be audio recorded. The interview will take approximately 30 minutes.

Participation in this study is entirely voluntary. You may ask the researcher to stop the interview at any time during the interview and you may withdraw from the study.

We are aiming to recruit 10 people living with MND and 10 of their carers in total.

The researchers do not expect your participation to cause any harm or discomfort to you.
However, if you experience feelings of distress as a result of participation in this study, please let the research team know immediately. You can also contact the following services for support:

- Lifeline – 13 11 14, www.lifeline.org.au
- Beyond Blue – 1300 22 4636, www.beyondblue.org.au
- MND SA - www.mndsa.org.au

If you feel uncomfortable at any time during an interview, you can ask the researcher to stop the interview or to take a break.

There are no costs to you if you participate in this research project.

Withdrawal Rights

You may, without any penalty, decline to take part in this research study. If you decide to take part and later change your mind, you may, without any penalty, withdraw at any time without providing an explanation. To withdraw, please contact the researcher. You may also refuse to answer any questions, and/or ask the researcher to leave.

If you do withdraw, any information that hasn't been included in data analysis can be withdrawn, but the information that has been included in analysis cannot be withdrawn. You should be aware that any information that is included in the data analysis will form part of the project results.

Confidentiality and Privacy

Only researchers listed on this form have access to the individual information provided by you. Privacy and confidentiality will be assured at all times. The research outcomes may be presented at conferences, written up for publication or used for other research purposes as described in this information form. However, the privacy and confidentiality of individuals will be protected at all times. You will not be named, and your individual information will not be identifiable in any research products without your

explicit consent. Once the research project ends, you will not be contacted by researchers unless you have given express permission for them to do so.

No data, including identifiable, non-identifiable and de-identified datasets, will be shared or used in future research projects without your explicit consent.

Data Storage

The information collected may be stored securely on a password protected computer and/or Flinders University server throughout the study. Any identifiable data will be de-identified for data storage purposes unless indicated otherwise. All data will be securely transferred to and stored at Flinders University for at least five years after publication of the results. Following the required data storage period, all data will be securely destroyed according to university protocols.

How will I receive feedback?

On project completion, a short summary of the outcomes will be provided to all participants via email or published on Flinders University's website. We anticipate the research information will be analysed by May 2023. You are welcome to contact the researchers at any time during and after the study if you have any questions regarding it.

Ethics Committee Approval
The project has been approved by Flinders University's Human Research Ethics Committee. Project number 4660.

Queries and Concerns

Queries or concerns regarding the research can be directed to Rebecca Francis on 08 82012811 or via email rebecca.francis@flinders.edu.au

If you have any complaints or reservations about the ethical conduct of this study, you may contact the Flinders University's Research Ethics & Compliance Office team via telephone 08 8201 3116 or email human.researchethics@flinders.edu.au.

Thank you for taking the time to read this information sheet which is yours to keep. If you accept our invitation to be involved, please sign the enclosed Consent Form.

CONSENT FORM – PARTICIPANT

Consent Statement

☐ I have read and understood the information about the research, and I understand I am being asked to provide informed consent to participate in this research study. I understand that I can contact the research team if I have further questions about this research study.

☐ I am not aware of any condition that would prevent my participation, and I agree to participate in this project.

☐ I understand that I am free to withdraw at any time during the study.

☐ I understand that I can contact Flinders University's Research Ethics & Compliance Office if I have any complaints or reservations about the ethical conduct of this study.

☐ I understand that my involvement is confidential, and that the information collected may be published. I understand that I will not be identified in any research products.

I further consent to:

☐ having my information audio recorded
☐ being contacted about other research projects

Signed:

Name:

Date: / /

Appendix 13.

Carer Information sheet and consent form. (Chapter 8)

INFORMATION SHEET AND CONSENT FORM – CARER

How do people living with Motor Neurone Disease and their primary carers learn about, understand and manage cognitive and behavioural change?

Chief Investigator
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Associate Professor Sebastian Doeltgen
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Tel: 08 7221 8817

Research Supervisor
Associate Professor Stacie Attrill
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Tel: 08 83133518

Description of the study

Chief Investigator Rebecca Francis is a PhD candidate at Flinders University, and this study will form part of her doctoral research program. We are inviting you to consider participating in a research project, which will explore your experiences of learning about, receiving education on, and if applicable, managing cognitive and behavioural change that can be a symptom of Motor Neuron Disease.

This Participant Information Sheet/Consent Form tells you about the study. It explains how the research will be done. Knowing what is involved will help you decide if you want to take part in the research.

Please read this information carefully. Ask questions about anything that you don't understand or want to know more about. Before deciding whether or not to take part, you might want to talk about it with a relative, friend or your local doctor.

Participation in this research is voluntary. If you don't wish to take part, you don't have to. You, or the person you care for, medical care, relationship with the university and/or the services which are received will not be affected by your decision.

If you decide you want to take part in the research project, you will be asked to sign the consent section. By signing it you are telling us that you:

- Understand what you have read
- Consent to take part in the research project
- Consent to participating in the screening assessment that is described

You will be given a copy of this Participant Information and Consent Form to keep.

This project is supported by Flinders University, College of Nursing and Health Science.

Purpose of the study

Some people living with Motor Neuron Disease may experience changes in their cognition, such as changes in thinking and problem-solving skills and/or their behaviour. Currently there is no information that tells us how people living with Motor Neurone Disease and their carers learn about this symptom of the disease, what education they receive, and how they are supported to manage these symptoms. It is also unclear how these symptoms may impact on how plwMND and their families manage swallowing difficulties. This research will explore your experiences related these symptoms.

We will do this by conducting semi-structured interviews with people who have MND and/or with people who care for someone who has MND. We anticipate that this interview will take approximately thirty minutes. Please note you can decide at any point, including during the interview to withdraw from the research or discontinue the interview.

During the interview, the lead researcher will ask you some general questions about your experience with MND, questions related to the symptom of cognitive and behavioural change and questions about the swallowing function and eating and drinking of the person with MND.

Benefits of the study

By sharing your experiences, you will help us understand how plwMND and their carers learn about symptoms of the disease, what educational resources they receive and how they like to receive this information. In addition, this research will help us understand how swallowing is managed every day, and if cognitive changes impact on swallowing management for both the plwMND and their primary carer.

By collecting this information, we will be able to improve health services for plwMND and their families. It will also provide education to health practitioners to assist them in making decisions and to help them to improve the care they provide.

We cannot guarantee or promise that there are any direct benefits to you from this research, however possible benefits may include more help for people to manage and make decisions about their cognitive and swallowing management in the future. There is little information about how people manage their swallowing during the course of MND, and this study will provide important information about this.

Recognition of Contribution / Time / Travel costs

If you would like to participate, in recognition of your contribution and participation time, you will be provided with a $40.00 Coles Myer gift card. This gift card will be posted to you on completion of the interview.

Participant involvement and potential risks

A member of the research team will give you information about the study. If you choose to participate in the study, you will need to sign a consent form. If you choose not to participate, you will not be contacted again by this research team about this study.

If you consent to participate in this study, you will be invited to attend an online interview via Microsoft Teams or Zoom or on the telephone. Alternatively, if it is your preference, a one-on-one face to face interview can be arranged at a location that is convenient to you. All interviews will be audio recorded. The interview will take approximately 30 minutes.

Participation in this study is entirely voluntary. You may ask the researcher to stop the interview at any time during the interview and you may withdraw from the study.

We are aiming to recruit 10 people living with MND and 10 of their carers in total.

The researchers do not expect your participation to cause any harm or discomfort to you.
However, if you experience feelings of distress as a result of participation in this study, please let the research team know immediately. You can also contact the following services for support:

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There are no costs to you if you participate in this research project.

Withdrawal Rights

You may, without any penalty, decline to take part in this research study. If you decide to take part and later change your mind, you may, without any penalty, withdraw at any time without providing an explanation. To withdraw, please contact the researcher. You may also refuse to answer any questions, and/or ask the researcher to leave.

If you do withdraw, any information that hasn't been included in data analysis can be withdrawn, but the information that has been included in analysis cannot be withdrawn. You should be aware that any information that is included in the data analysis will form part of the project results.

Confidentiality and Privacy

Only researchers listed on this form have access to the individual information provided by you. Privacy and confidentiality will be assured at all times. The research outcomes may be presented at conferences, written up for publication or used for other research purposes as described in this information form. However, the privacy and confidentiality of individuals will be protected at all times. You will not be

named, and your individual information will not be identifiable in any research products without your explicit consent. Once the research project ends, you will not be contacted by researchers unless you have given express permission for them to do so.

No data, including identifiable, non-identifiable and de-identified datasets, will be shared or used in future research projects without your explicit consent.

Data Storage

The information collected may be stored securely on a password protected computer and/or Flinders University server throughout the study. Any identifiable data will be de-identified for data storage purposes unless indicated otherwise. All data will be securely transferred to and stored at Flinders University for at least five years after publication of the results. Following the required data storage period, all data will be securely destroyed according to university protocols.

How will I receive feedback?

On project completion, a short summary of the outcomes will be provided to all participants via email or published on Flinders University's website. We anticipate the research information will be analysed by May 2023. You are welcome to contact the researchers at any time during and after the study if you have any questions regarding it.

Ethics Committee Approval
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Queries and Concerns

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Thank you for taking the time to read this information sheet which is yours to keep. If you accept our invitation to be involved, please sign the enclosed Consent Form.

CONSENT FORM – PARTICIPANT

Consent Statement

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☐ I am not aware of any condition that would prevent my participation, and I agree to participate in this project.

☐ I understand that I am free to withdraw at any time during the study.

☐ I understand that I can contact Flinders University's Research Ethics & Compliance Office if I have any complaints or reservations about the ethical conduct of this study.

☐ I understand that my involvement is confidential, and that the information collected may be published. I understand that I will not be identified in any research products.

I further consent to:

☐ having my information audio recorded
☐ being contacted about other research projects

Signed:

Name:

Date: / /

Appendix 14.

What matters to you? My Goals of Care Planner. SALHN (2024) (Chapter 10).

Image removed due to copyright

Appendix 15.

Research dissemination. MND Association newsletter (Chapter 8)