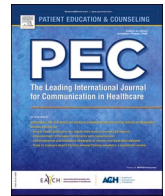




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## Exploring clinical management of cognitive and behavioural deficits in MND. A scoping review

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## ABSTRACT

**Objectives:** Little is known about how cognitive and behavioural decline in MND is managed clinically. This review aimed to summarise clinical management approaches of cognitive and behavioural decline in MND reported in peer-reviewed and grey literature.

**Methods:** A scoping review was conducted across Embase, Medline, Psychinfo and Emcare in October 2022. Grey literature was also searched across Google Scholar and Google in October 2022. **Results:** A total of N = 26 studies and 8 documents were included. Thematic analysis revealed six key areas of clinical management: i. Assessment, ii. Education, iii. Advance Care Planning, iv. Adaptation of Care Plan, v. Communication and vi. Carer Support. **Conclusions:** The literature on management of cognitive and behavioural decline in MND is sparse. Most peer-reviewed literature consists of expert commentary and there is a lack of primary data to guide practitioners and families on how to manage cognitive and behavioural change in MND. **Practice Implications:** Determining as early as practicable the presence of cognitive and behavioural changes in pwMND will enable practitioners to make adaptations to communication, provide education and supported decision-making for forward planning. This will enable individualised care, planned in partnership with families with MND, which incorporates personal needs and wishes.

## 1. Introduction

Motor Neurone Disease (MND) is an umbrella term for a class of heterogenous neurodegenerative terminal diseases, of which the most common is amyotrophic lateral sclerosis (ALS) [1]. Although previously thought to be only associated with degeneration of primary motor cortical circuits [2,3], MND is increasingly recognised as a multi-system disease [2,4], associated with changes to cognition and behaviour [3,5]. Clinically, cognitive and behavioural deficits are linked to poorer patient outcomes [6], reduced treatment uptake [7], reduced adherence [7,8] and shortened life expectancy [9,10,11]. In addition, these symptoms have been shown to increase caregiver burden [8,12,13]. These negative impacts are important to consider in management, as cognitive and behavioural decline associated with MND is thought to occur in up to 50 % of people with the disease (pwMND), of which 35 % experience

mild-moderate symptoms, and 15 % will meet the diagnostic criteria for fronto-temporal dementia [14,15,16].

Literature referring to cognitive and behavioural decline 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 [9,17], whereas behavioural deficits may manifest as apathy, rigidity and disinhibition [18]. Current clinical guidelines for MND recommend routine care encompasses consideration of capacity, screening for cognitive and behavioural decline, attaining informed consent, and healthcare professional training for cognitive changes [19,20,21]. In some instances, complete neuropsychological assessment is necessary, with further linkage and reference to dementia specific guidance [21]. However, these guidelines do not extend to specific recommendations or strategies

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to support pwMND who experience cognitive and behavioural symptoms, instead providing general considerations relating to: tailoring care, communication, advanced decision-making and end of life planning.

Early identification of cognitive and behavioural deficits, regular ongoing assessment, and specific strategies for supporting practitioners who provide care, and pwMND and their families are critical to facilitate understanding of the disease and improve the lived experience of families. This is particularly important in the context of person-centred care [22], where care planning is conducted in partnership with pwMND and their families, and management respects and incorporates personal needs and wishes. For pwMND, cognitive and behavioural skills influence 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 practitioners, 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 [23,24], limited evidence exists regarding the specific strategies that may guide practitioners to manage cognitive and behavioural decline once identified. This may lead to practitioners, 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 care-planning [25]. PwMND concurrently present with progressive communication impairment due to 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. 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, families and practitioners, a scoping review was conducted to identify and collate strategies specific to managing cognitive and behavioural decline in MND, addressing the following research question:

*“How is cognitive and behavioural decline associated with MND managed?”.*

## 2. Methods

We conducted a Scoping Review using the Arksey and O'Malley [26] framework, which allowed exploration of peer-reviewed and grey literature, using a rigorous process to identify and map evidence [27]. This approach also allowed the inclusion of clinical guidelines, MND support services information, patient information sheets and similar. It was important to capture the grey literature accessible to pwMND, their families and practitioners to allow analysis of the full range of information and resources available to guide clinical practice [28].

A medical librarian assisted with designing the master search. Initial search terms related to MND AND cognition & behaviour AND management (Table 1) were included. The search was restricted to English and a publication date from 2010 onwards to capture contemporary care approaches to intervention, 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 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, Psycinfo and Emcare in October 2022. Further

snowball searching of the reference lists of included studies resulted in one additional study [20] identified for inclusion. The search was translated and run in Google Scholar and Google in October 2022 to access the relevant grey literature.

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 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 (see Fig. 1).

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 duplicates and removed. Two reviewers, 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 [29], including country of publication, publication year and study type as represented in Fig. 2. Additionally, a verbatim copy of text that directly related to the research question and search terms was extracted.

Two members of the research team completed analysis of the extracted qualitative data related to the management of cognitive and behavioural decline. 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 [30]. 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.

## 3. Results

Of the 26 included peer-reviewed studies (Fig. 2), nine were original research with primary data. Of these, five were research studies, which included a four-round Delphi model with practitioners working with pwMND [31], one cross sectional cohort study of 51 caregivers of pwMND [32], one cross sectional cohort study of 15 caregivers of pwMND [33], a mixed methods study of 22 ALS care services [34] and a factor analysis study of 39 pwMND with cognitive and behavioural change [32]. 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 Fig. 2.

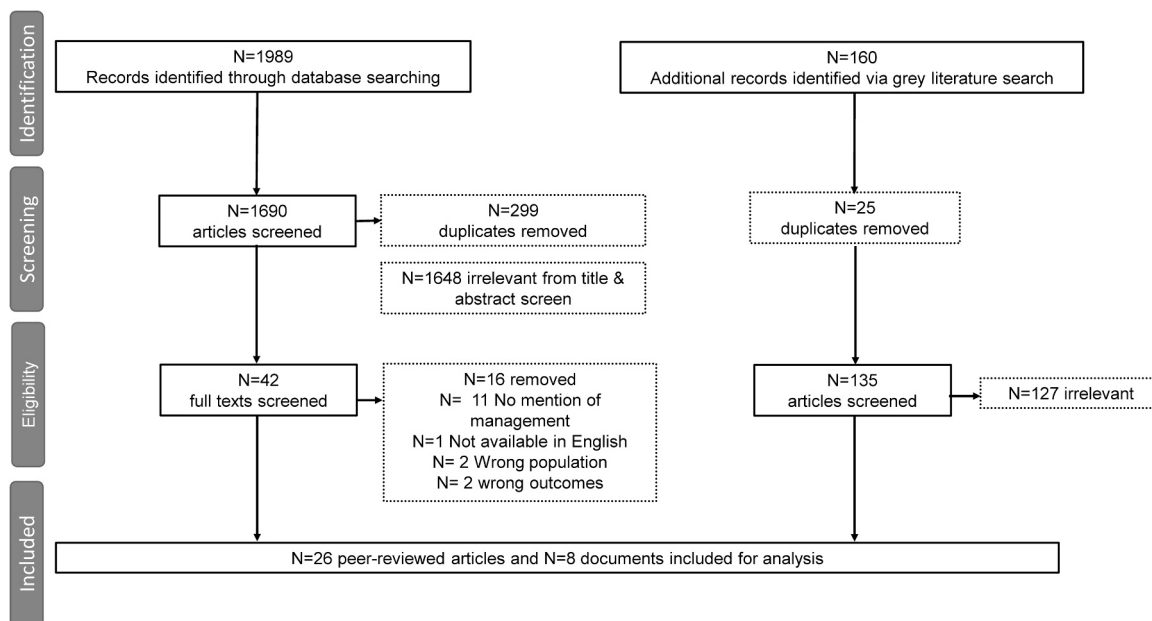


Fig. 1. TIFF PRISMA flow diagram.

### Data charting

Peer review N = 26			
Original Research Primary Data		Non Primary Data	
Research study	<ul style="list-style-type: none"> <li>Caga, 2018 AUS</li> <li>Caga, 2021 AUS</li> <li>Olesen, 2022 EU</li> </ul>	<ul style="list-style-type: none"> <li>Hodgins, 2020 UK</li> <li>Radakovic, 2020 UK</li> </ul>	<ul style="list-style-type: none"> <li>Miller, 2009 USA</li> <li>Lee, 2012 HKG</li> <li>++Clare, 2019 UK</li> <li>van Eenennaam, 2020 UK</li> </ul>
Dissertation	<ul style="list-style-type: none"> <li>Bertolin, 2018 USA</li> <li>Crockford, 2018 UK</li> </ul>		
Case study	<ul style="list-style-type: none"> <li>Bialkowska, 2018 EU</li> </ul>		
Audit	<ul style="list-style-type: none"> <li>Stavrou, 2020 UK</li> </ul>		
++ Paper reported on neurodegenerative conditions.			
Other			
Review Expert commentary	<ul style="list-style-type: none"> <li>Jenkins, 2014 UK</li> <li>Fried-Oken, 2015 USA</li> <li>Houseman, 2015 USA</li> <li>Khin Khin, 2015 USA</li> <li>Oliver, 2016 UK</li> <li>Danel-Brunaud, 2017 EU</li> <li>Hodgen, 2017 AUS</li> <li>Everett, 2020 USA</li> <li>Caga, 2019 AUS</li> </ul>	<ul style="list-style-type: none"> <li>Oliver, 2017 UK</li> <li>Shoesmith, 2020 CAN</li> </ul>	<ul style="list-style-type: none"> <li>Lugassy, 2016 USA</li> </ul>
	<ul style="list-style-type: none"> <li>Guidelines</li> </ul>		<ul style="list-style-type: none"> <li>Training material (Abstract)</li> <li>Lindenberger, 2013 USA</li> </ul>
		<ul style="list-style-type: none"> <li>Educational Material Organisation Resource</li> </ul>	
Countries Tally			
United Kingdom = 9, USA = 8, European Union = 4, Australia = 3, Canada = 1, Hong Kong = 1.			

Fig. 2. TIFF Data charting.

The eight included grey literature documents (Table 2) 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 [21], 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 [35] 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 practitioners and consumers.

**Table 1**

Theme 1. Cognition and behaviour assessment.

Citation	Strategy	Management description	Enacted by	Recipient
Khin Khin, 2015	SS	<b>“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 behavioral 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 2–5 min, a more extensive assessment of 5–20 min, and formal neuropsychiatric testing. As they noted, screening tests cannot be used in formal diagnosis of cognitive ALS-FTD, ALSbi, or ALSci”. Pg. 212</b> <b>“By conducting brief cognitive and behavioral screening of patients at regular intervals, those who start to show deficits in these areas can be identified and monitored closely”. Pg. 216</b>	Practitioner	pwMND
Stavrou, 2020	SS	<b>“Additional pathways should be developed for cognitive/behavioral 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</b>	Practitioner	Practitioner
van Eenennaam, 2020	SS	<b>“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 Behavioral ALS Screen can be used to gain insight into affected cognitive domains.” Table 1. Pg 5</b>	Practitioner	pwMND
Shoesmith, 2020	GR	• Screening for cognitive and behavioural impairment should be performed in patients with ALS early in their disease (level B). • 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	Practitioner	pwMND
Crockford, 2018	GR	“Recently updated ALS consensus guidelines suggest 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	Practitioner	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 behavioral impairment is an essential component of clinical care for this population” pg.54	Practitioner	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	Practitioner	pwMND
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	Practitioner	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	Practitioner	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	Practitioner	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	Practitioner	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	Practitioner	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.

Thematic analysis of the included literature revealed six distinct themes that described a specific feature of cognitive and behavioural decline in MND to manage (Table 1–6). 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.

### 3.1. Theme 1 Cognitive and behaviour assessment

Screening for cognitive and behavioural deficits in MND was discussed in 12 of the 26 included studies, 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, fronto-temporal dementia and MND” published by the MND Association for the

MND Community of Practice [36] 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 [21] 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 [21] 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” [37] and the “ALS Cognitive Behavioural Screen (ALS-CBS)”

**Table 2**  
Theme 2. Education.

Citation	Strategy	Management Description	Enacted by	Recipient
Radakovic, 2020	SS	<b>“...educational material on common cognitive and behavioral 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</b> <b>“Information/education for caregivers/families and staff about impairment and impact” Pg. 21</b>	Practitioner	Practitioner & Caregiver
Danel-Brunaud, 2017	SS	<b>“Education and counselling on the neurological bases of behavioral 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</b>	Practitioner	Caregiver
Houseman, 2015	SS	<b>“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</b> <b>“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</b> <b>“Address (possible) behavioral 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 utilizing soothing activities”. Pg. 126</b>	Practitioner	Caregiver
Caga, 2021	GR (Derived from literature about populations with dementia).	<b>“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</b> <b>“...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</b> <b>Other approaches were more personalized in terms of providing more general support, education and information for family members or caregivers about cognitive and behavioral impairment, which are also often observed in management of dementia, for example”. Pg. 21–22</b>	Practitioner	pwMND & Caregiver
Radakovic, 2020	GR (Derived from literature about populations with dementia).	<b>“Expert consensus is that care provision in ALS should include interventions directed toward the management of cognitive and behavioral 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 behavioral impairment.” Pg. 211</b> <b>“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</b>	Practitioner	Caregiver
Hogden, 2017	GR	<b>“In dementia, education focusing on training caregivers to modify their interactions with patients appears to be highly beneficial for managing behavioral symptoms” Pg. 603</b>	Practitioner	Caregiver
Van Eenennaam, 2020	GR		Practitioner	Caregiver
Caga, 2018	GR (Derived from literature about populations with dementia).		Practitioner	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.

[23] which are both validated for use with this population. Four of the included peer-reviewed studies [20,38] 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 decline 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.

### 3.2. 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 [39,40] 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 [31] suggested that education should include common presentations of cognitive and behavioural change in MND to support practitioners 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 practitioners (Table 4). Of note, two studies referred to dementia-related literature [31,41] 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 [36] (pg. 9), practitioner 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 [41].

### 3.3. Theme 3 Advanced care planning

Five studies (Table 5) 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

Table 3

Theme 3. Advanced care planning.

Citation	Strategy	Management description	Enacted by	Recipient
Everett, 2020	SS	<b>“ACP in ALS should start early...and should continue throughout the disease course as treatment preferences often change with symptom progression. Many clinicians 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</b>	Not provided	Not provided
Houseman, 2015	SS	<b>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</b>	Practitioner	pwMND
Shoesmith, 2020	GR	“Because the presence of frontotemporal dementia negatively affects survival, ACP should be done early in the disease (EC).” <b>Table 1. Pg. E1458</b>	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	Practitioner	pwMND
Khin 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 honored 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.

families as part of routine practice, as cognitive impairment is common in ALS. The authors noted that this approach may address concerns related to the sensitivity of the topic, and further suggested that families welcome these discussions [42]. 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 5) 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 [43] 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 [36] is described (page 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.

### 3.4. 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 practitioners, including specific strategies to manage deficits: using a supportive communication style, addressing safety concerns, supporting decision-making and adapting the environment (see Table 6 for details). The remaining studies provided *general recommendations* that the care plan should be adapted or adjusted but without describing specific information (Table 6).

The grey literature provided more *specific strategies* related to adapting care than the peer-reviewed literature. Specifically, the MND Association guide (pages 27–35) [36] outlines strategies for practitioners 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 [21] 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 [21] 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 [19] 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 practitioners, it is also noted that these resources draw from evidence generated from outside of the MND context.

### 3.5. 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 5), one provided *specific strategies* to support practitioners to communicate with pwMND by delivering information compassionately and with sufficient time to allow the pwMND to process the information [31].

In the remaining five studies (Table 5), the *general recommendations* made suggestions that communication can be difficult with a pwMND who has cognitive and behavioural changes and, therefore, communication should be adjusted. However, specific strategies to guide practitioners were not discussed. One way of adjusting communication was described as using alternative and augmented communication (AAC) to support impaired communication (Table 5); However, cognitive decline was also suggested to impact the use and uptake of such communication tools [44,45].

### 3.6. 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 6), of which two described *specific strategies* addressing support for carers to manage these changes. An intervention program called “Embrace” [33] described cognitive and behaviour support aimed at improving the experience for

**Table 4**  
Theme 4. Adaptation of care.

Citation	Strategy	Management description	Enacted by	Recipient
Radakovic, 2020	SS (Derived from literature about populations with dementia).	<b>Structured support approaches</b> (Taken from Table 2) Pg. 21 <b>Strategies for managing the deficits or issues identified:</b> <ul style="list-style-type: none"> <li>• <b>Adapting the environment</b> (e.g., more/increasing structure, less distraction, safety measures)</li> <li>• <b>Breakdown tasks into small manageable steps</b></li> <li>• <b>Verbal interaction</b></li> <li>• <b>Distraction</b></li> <li>• <b>Routine</b></li> <li>• <b>Physical focus</b></li> <li>• <b>Behavioral approach</b></li> <li>• <b>Person centered.</b></li> <li>• <b>Family work/systemic approach</b></li> <li>• <b>Acceptance and commitment therapy</b></li> </ul> <b>Personal support approaches</b> <ul style="list-style-type: none"> <li>• <b>Compassionate communication</b></li> <li>• <b>Increased time</b></li> <li>• <b>Make suggestions and let person come to own conclusions</b></li> </ul> “...evidence-based techniques, such as cognitive stimulation therapy and behavioral 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 standardized practice or guidance in relation to interventions”. Pg. 22	Practitioner	Practitioner 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	Practitioner Caregiver	pwMND
Oliver, 2016	SS	“...simple decision between two clear options to facilitate the patient’s involvement” Pg. 67	Practitioner Caregiver	pwMND
Clare, 2019	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	Practitioner	Not detailed
Stavrou, 2020	GR	“care planning should be adapted for people with cognitive impairment”. Pg. 460	Practitioner	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	Practitioner	pwMND
Miller, 2009	GR	“There are insufficient data to support or refute the impact of cognitive and behavioral 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.

carers of pwMND. The second study [31] 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 6), however, without providing specific strategies to guide practitioners to enact this.

The theme of Carer Support also appeared in the grey literature, with the MND Association Guide [36] 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 [21] 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 behavioural change was recognised in both the peer-reviewed and grey literature. A small number of resources are available to guide practitioners 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.

## 4. Discussion and conclusion

### 4.1. Discussion

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 practitioners, carers, pwMND and researchers. These themes

**Table 5**  
Theme 5. Communication.

Citation	Strategy	Management description	Enacted by	Provided to
Radakovic, 2020	SS	<b>(Taken from Table 2) Pg. 21</b> - <i>Compassionate communication</i> - <i>Increased time</i> - <i>Make suggestions and let person come to own conclusions</i> “...breaking down tasks into manageable small steps, verbal interaction and routine were noted as the best approaches”. Pg. 19	Practitioner	pwMND
Shoesmith, 2020	GR	<ul style="list-style-type: none"> <li>• <i>Simplify decision-making (e.g., limit choices and reduce open-ended questions)</i></li> <li>• <i>Support problem-solving (e.g., specify the topic being discussed, prompt if needed)</i></li> <li>• <i>Clarify complex information (e.g., breakdown information, explain terminology that is unfamiliar) Pg. 19</i></li> </ul> “The choice of communication devices should be tailored to the patient’s needs and abilities”. “Patients with cognitive impairment may need individualized 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	Practitioner	pwMND
Hodgins, 2020	GR	“...facilitating communication between family members to promote understanding and manage stressful situations...” Pg. 97	Practitioner	pwMND & Caregiver
Everett, 2020	GR	“...behavioral changes make communication difficult even if patients maintain decision-making capacity”. Pg. 843	Practitioner	pwMND
Oliver, 2017	GR	“Tailor discussions to the person’s needs, taking into account their communication ability, cognitive status and mental capacity” Pg.320	Practitioner	pwMND
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	Practitioner	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.

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:

- i. the use of the ECAS or ALS CBS for early screening and as part of regular and ongoing management.
- ii. simplifying communication with people identified to have cognitive and behaviour change to support understanding.
- iii. early instigation of advanced care planning to allow for changes in capacity.

iv. providing education which focusses on the neurological basis behind the functional changes.

v. 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 decline in MND is derived from descriptive

**Table 6**  
Theme 6. Carer Support.

Citation	Strategy	Management description	Enacted by	Recipient
Olesen, 2022	SS	<b>“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</b> “...new online palliative rehabilitation program (EMBRACE), a blended learning program developed for caregivers of PALS/Cis.” Pg. 2	Practitioner	Caregiver
Radakovic, 2020	SS	<b>“Support the family/caregiver (e.g., support the family/caregiver in understanding cognitive and behavioral symptoms, expressing themselves to the patient, taking respite).” pg. 19</b>	Practitioner	Caregiver
Caga, 2019	GR	“Comprehensive management of cognitive and behavioral symptoms not only promotes holistic care of patients but would also further enhance caregiver’s psychological well-being...” Pg. 4	Practitioner	pwMND
Hodgins, 2020	GR	“Interventions mentioned by psychologists included; behavioral 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	Practitioner	Caregiver
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	Practitioner	Caregiver
Hogden, 2017	GR	“Expert consensus is that care provision in ALS should include interventions directed toward the management of cognitive and behavioral impairment in ALS, including... support services for family carers on how best to support and care for ALS patients who have cognitive or behavioral impairment. In this context neuropsychologists make an important contribution to the care of ALS patients and their caregivers” Pg. 211	Practitioner	Caregiver
Danel-Brunaud, 2017	GR	“Education and counselling on the neurological bases of behavioral 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	Practitioner	Caregiver
Caga, 2018	GR (Derived from literature about populations with dementia).	“... strategies for ALS caregivers to manage apathy include cognitive behavioral strategies to ensure realistic expectations about a patient’s capacity to engage in activities as well as participation in alternative activities” Pg. 603	Practitioner	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.



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 [46] 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 which excludes pwMND with cognitive and behavioural changes. As up to fifty percent of pwMND experience changes to cognition [47], existing research is potentially lacking validity for a large proportion of the MND population [47]. As these symptoms are known to result in poorer intervention outcomes and prognosis for pwMND [48,49], 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, practitioners 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 [50] or gastrostomy to manage nutritional and fluid intake [51]. 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 practitioners 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 practitioners 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 [21,52] 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 [53,54]. 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 may 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) [55] 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 practitioner 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 [21] 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 practitioners that ultimately improves the understanding and experiences of affected individuals and their families. Practitioners 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 interfaces, 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 to better understand the attitudes of both practitioners and families with MND towards cognitive and behavioural supports, including any barriers that may exist to accessing these [56]. As highlighted in the NICE guidelines [21], it is not yet understood if available services are accessible, accessible, or if they improve outcomes as anticipated.

#### 4.2. Conclusion

The literature on management of cognitive and behavioural decline 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 practitioners 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 decline. 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 [21] 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.

#### 4.3. 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 practitioners 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.

#### CRedit authorship contribution statement

**Francis Rebecca:** Writing – review & editing, Writing – original draft, Project administration, Methodology, Investigation, Formal analysis, Data curation, Conceptualization.

#### Declaration of Competing Interest

There are NIL competing interests.

#### Appendix A. Supporting information

Supplementary data associated with this article can be found in the online version at doi: 10.1016/j.pec.2023.107942.

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