MND Association | For professionals

# Cognitive change and frontotemporal dementia in MND

# About Motor Neurone Disease

**Today, six people**

will be told they have MND and six people will die from the disease. There is no cure.

**MND is a fatal, rapidly progressing disease.**

It affects the nerves in the brain and spinal cord that control muscles.

**A third of people will die within a year of diagnosis.**

More than half will die within two years.

**People may lose movement, speech, swallowing and breathing.**

This affects quality of life for them and those around them.

**MND doesn't discriminate.**

It affects people from all backgrounds and at all ages.

**People with MND may experience changes in thinking and behaviour.**

Some develop frontotemporal dementia, with more severe changes.

**MND affects everyone differently.**

Symptoms progress at different speeds and a different order for everyone.

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# Introduction

Motor Neurone Disease (MND) is a progressive, fatal disease that attacks the motor neurones, or nerves, in the brain and spinal cord. It leads to muscle weakness and can cause difficulties with moving, breathing, swallowing and speaking. About two people in every 100,000 people in the general population will develop MND each year.[1]

## An overview of cognition and behaviour in MND

MND also includes non-motor symptoms. One of the most common is cognitive change.[2] The term "cognition" refers to high-level brain functions that can be categorised into different areas:

- executive functioning, such as planning or inhibiting responses
- language, including understanding language, speaking or writing
- behaviour and social cognition, for instance understanding and interpreting other people's thoughts, beliefs and feelings
- memory, such as acquiring and remembering information
- perception, for example processing the information gathered by the five senses.

Research has found that people with MND can show changes in the frontal and temporal areas of the brain, affecting thinking, reasoning and behaviour. About 50% of all people with MND experience some cognitive or behavioural changes.[2] This figure rises to 80% in the final stages of the disease.[3]

Approximately 35% of those with MND experience mild impairments in executive functions, language and/or social cognition.[2,4] However, for others, the symptoms can be more pronounced.

People can be diagnosed with MNDci (MND with cognitive changes), MNDbi (MND with behavioural changes) or MNDcbi (MND with a combination of cognitive and behavioural changes).[5,6]

Up to 15% of individuals can also be diagnosed with frontotemporal dementia (FTD).[2] This usually affects people below the age of 65, and it presents different signs and symptoms than more common forms of dementia, such as Alzheimer's disease.[4,7]

People with FTD often experience severe changes in behaviour and executive functions. They can also present with changes in language or conceptual understanding.

FTD can be diagnosed before, at the same time, or after MND. Moreover, up to 15% of people initially diagnosed with FTD go on to develop MND.[8] In these cases, dementia is identified before movement is impaired and MND is diagnosed.

**Note:** While these figures are based on current evidence, the number of people with MND affected by cognitive or behavioural changes may be higher. Numbers may vary depending on the sensitivity of the tool used to detect these symptoms and the stage of the disease.[5]

## About this guide

This booklet contains information on cognitive and behavioural changes and FTD. It has been designed to help health and social care professionals support individuals with MND experiencing these symptoms.

This guide is split into three sections:

**Section 1: Understanding cognition and behaviour in MND** summarises cognitive and behavioural symptoms observed in people with this condition.

**Section 2: Assessing cognitive and behavioural changes** includes tools for screening people with MND for these symptoms.

**Section 3: A practical toolkit** helps professionals work together to support the people with MND, their carers and families.

# 1

# Understanding cognition and behaviour in MND

# Changes in cognition and behaviour

People with MND can experience deficits in executive functions, language and/or social cognition.

Executive impairments are common as they heavily depend on the brain's frontal lobes. However, language changes can be equally common and may occur without executive dysfunction.[6] Behavioural changes, such as apathy and disinhibition, can also be experienced with or without cognitive changes.

International diagnostic criteria established that MNDci can be diagnosed in various ways:[6]

- when there's evidence of impaired verbal fluency (see page 9)
- when there are two or more impaired executive domains (including social cognition)
- when there are language changes
- when there's a combination of executive and language deficits.

MNDbi is diagnosed based on evidence of apathy or at least two of the symptoms listed on pages 11–12.[6] People with MNDbci meet the criteria for both MNDci and MNDbi, but the symptoms are not severe enough for a diagnosis of FTD.[6]

See **pubmed.ncbi.nlm.nih.gov/28054827/** to find out more about diagnostic criteria.

## Executive dysfunction

Executive functions help set and achieve goals, monitor performance and maintain attention. They also include cognitive flexibility, the ability to deduce rules, and the ability to adapt to change and feedback.[6]

| Issues caused by executive dysfunction: | Examples of everyday life activities affected: |
|---|---|
| multitasking, timekeeping and organising | managing affairs and finances |
| concentrating, maintaining attention and shifting focus to other things | doing more than one thing at a time, for example ironing while watching TV |
| inhibiting or controlling thoughts | holding conversations if there are background noises |
| making decisions, setting goals or making and implementing a plan | planning for the future or making care decisions |
| organising and monitoring tasks | completing work |
| sequencing tasks | cooking a meal or getting ready in the morning |
| generating ideas, thinking flexibly and finding solutions | adapting to MND or other conditions |
| understanding or deducing rules and instructions. | starting new activities or learning new tasks. |

> **“People can have planning and organising issues. For example, making a meal is a multi-step process: chop, season, use the oven. Often, people get really worked up trying to plan and organise that. This can lead to problems, especially if they’re living alone.”**
>
> MND professional

## Language dysfunction

Understanding whether language issues are caused by cognitive changes or speech difficulties can be challenging.

Up to 80% of people with MND experience speech problems (dysarthria) due to muscle weakness in the mouth, throat and face. These people may rely on alternative and augmentative communication (AAC) aids or lose the ability to speak, which could mask the language deficits caused by changes in brain functions.

Verbal fluency is often assessed to identify executive dysfunction.[6] Language deficits can also arise independently from impaired executive functions.

Verbal fluency is the ability to quickly generate words that start with a specific letter (letter fluency) or belong to a particular semantic category (such as a list of animals). It is commonly impaired in people with MND with cognitive changes, although it may be more pronounced in those affected by pseudobulbar palsy (emotional lability, see page 30).

Some people with MND may also show severe primary language impairment, known as aphasia (see page 13).

Language deficits include:[6]

- difficulties quickly generating or coming up with words (especially verbs)
- reduced verbal expression and initiation – for example using shorter sentences
- problems with spelling or grammar, such as not completing sentences, missing determinants such as “the” and “an”, or misusing verbs
- repeating words and phrases no longer appropriate or relevant to the situation

- using stereotyped expressions
- copying parts of what someone else just said (echolalia)
- difficulties finding words while talking – for instance, the person pausing to search for a word or talking around it (circumlocution)
- misunderstanding complicated sentences
- problems understanding the meaning of words, especially verbs
- issues maintaining a logical flow or staying on topic while talking.

> **"He became very frustrated, as he thought his writing made sense, but it didn't."**
>
> Carer of a person with MND

## Changes in behaviour and social cognition[4–6]

People with MND may experience behavioural changes with or without cognitive impairments. However, it's unclear whether problems with social cognition are independent of executive deficits.[6]

Someone may seem to go through a personality change, becoming restless, irritable and, in some instances, aggressive. Long-standing traits may become more pronounced. For example, a determined person may now seem stubborn and inflexible.

People with deficits in social cognition also show altered emotional processing, reduced recognition of emotional facial expressions, especially negative ones, and impaired theory of mind.[4,6]

Theory of mind is the ability to understand other people's feelings, thoughts, beliefs and intentions.[5,9] Impairments lead to loss of sympathy and empathy, with reduced responsiveness to social cues and feelings of others (including their partner, carer, family or friends).

> **“I feel as though I am in conflict with another individual inside me, but not part of me. It’s as if an unknown force is taking possession of my body and gradually pushing me out.”**
>
> Person with MND

| **Behavioural problems:[6]** | **Examples:** |
|---|---|
| apathy and inertia | loss of interest in social situations or everyday activities |
| | issues with starting a task |
| | withdrawing from others and acting distant |
| loss of empathy and sympathy for others | being unable to respond to social cues |
| | not caring or noticing the needs and feelings of others (including relatives and friends) |
| disinhibition and loss of self-control | acting in socially inappropriate ways |
| | making disinhibited comments |
| | losing social manners |
| | impulsive, rash or careless actions, including gambling or inappropriately shopping online or from cold callers |

| Behavioural problems:[6] | Examples: |
|---|---|
| perseverative, rigid, stereotyped or compulsive, ritualistic behaviour | repeating simple movements |
| | hoarding items |
| | using the same phrases and responses repeatedly (for example a catchphrase) |
| | performing actions repeatedly like a ritual |
| changes in eating habits or hyperorality (tendency to put objects in the mouth to examine them). | compulsively overeating |
| | excessive licking or chewing |
| | cramming food |
| | developing new or altered preferences for certain foods (often sweet foods)[10] |
| | drinking or smoking too much or more than usual. |

> **"People may have impaired judgement. For example, they might want to cross a busy street without checking or go out with their shirt open."**
>
> MND professional

# Frontotemporal dementia (FTD)

People with MND and FTD (MNDFTD) experience MND symptoms alongside severe cognitive and behavioural changes typical of frontotemporal dementia.

There are various types of FTD, including:

**Behavioural variant FTD (bvFTD):** This is the most common presentation in people with MND. It is marked by significant personality shifts, executive dysfunction and behavioural changes such as apathy, disinhibition, perseveration, emotional bluntness and loss of insight.[7,8]

> **"People can become very egocentric, acting like they are only interested in themselves and being unable to understand others' emotions. This can be very hurtful to those around them."**
>
> MND professional

**Progressive non-fluent aphasia (PNFA):** Also known as the non-fluent variant of primary progressive aphasia (nfvPPA), PNFA affects a person's ability to produce and use words in spoken or written communication. This is known as aphasia. Features include:[7]

- reduced speech output
- grammatical errors when speaking or writing
- difficulties in recalling words (especially verbs)
- mispronouncing words, for example saying "pish" for "fish"
- difficulty understanding syntactically complex sentences, such as those using passive verbs, for example "the cat was chased by the dog".

**Semantic dementia (SD):**[7] This variant of FTD is also known as semantic variant PPA (svPPA). It's less common in people with MNDFTD, and it causes problems with understanding concepts. The key signs are:

- difficulty naming objects and understanding what words mean
- semantic errors, including mistaking the meaning of words (for instance, calling a tiger a "dog")
- fluent but empty speech
- difficulty recognising faces and objects.

Semantic problems are associated with the shrinkage of the brain's temporal lobes, which can be more pronounced on either the left or right side.

## MNDFTD diagnosis

There are three ways to diagnose MNDFTD:[6]

- progressive behavioural and/or cognition decline with at least three behavioural symptoms (see the table on pages 11–12)
- language impairments meeting the criteria for semantic dementia or PNFA. These may co-exist with other cognitive or behavioural symptoms
- two or more cognitive or behavioural symptoms, alongside loss of insight (lack of awareness of one's condition) and/or psychotic symptoms.

Psychotic symptoms can include delusions (such as the belief that someone is trying to harm them), hallucinations (for example hearing voices or feeling sensations on the skin) and, sometimes, paranoia, depression or mania. Although rare, these symptoms may occur, especially in those with mutations in the C9ORF72 gene.[11,12]

People with psychotic symptoms may experience greater problems with attention, memory, executive functions, disinhibition, apathy and stereotypical behaviours.[11,12] This can impact quality of life, lead to social isolation and increase carers' stress.

> **"Changes may come gradually, but carers or family members may tell you things like: 'He's being awkward' or 'He doesn't understand that I'm hurt'."**
>
> MND professional

When assessing symptoms to diagnose MNDFTD, consider the following:

- Changes should not be explained by the physical restrictions caused by MND.
- Apathy, inertia and lack of empathy should not be caused by depression, fatigue, emotional lability or respiratory issues (see pages 30–33).
- People may exhibit varying degrees of behavioural and cognitive changes. They may show significant alterations in social behaviour, while still performing relatively well on standard tests of executive function. In such cases, assessing emotion recognition can help detect early cognitive change.
- Cognitive and behavioural changes can occur at any stage of the disease. However, it's unclear whether FTD can develop when there are no or only mild signs of cognitive changes in the early stages.[5]

Research studies have produced mixed results regarding the relationship between the progression of MND, cognitive and behavioural symptoms, and FTD.[3,5,13,14]

Symptoms of FTD may appear before motor signs, masking them. For this reason, regular assessments are essential. Remember that people with MND can still develop other forms of dementia.[6]

Additionally, studies have found that people with FTD and C9ORF72 gene mutations are at a higher risk of developing MND and should therefore be closely monitored.[15]

Mutations in the C9ORF72 gene are the most common cause of inherited FTD, MNDFTD and familial MND (5-10% of all MND cases). They have also been associated with up to 10% of MND cases with no family history. Additionally, they have been linked to more severe cognitive deficits, even in people without dementia.[5]

Some studies also suggest that people with bvFTD with language problems, poor verbal fluency and apathy may be more vulnerable to developing MND and should be closely monitored.[16] Those with bvFTD+PNFA may also be at a higher risk of developing MND.[15]

# Other changes

## Changes in brain function and structure[3–5]

People experiencing cognitive and/or behavioural symptoms present changes in the brain's structure and function. These are usually more severe in those with MNDFTD.

Changes include alterations in blood flow and in the grey and white matter in the frontotemporal regions and the motor cortex.

Imaging studies also show shrinking in these brain regions, which may be used as a biomarker of cognitive decline.[4] However, it's essential to take into account age-related atrophy.[6] Post-mortem studies have also shown abnormal build-up of proteins like TDP-43 and loss of connections between nerve cells (synapses).[8,17,18]

## Changes in memory and visuospatial functions

People with MND may have issues recalling information, rather than forgetting it. However, difficulties maintaining attention due to executive dysfunction may also impact memory.[5]

Perceptual processes are not usually impaired in MND and FTD. Problems with spatial orientation and locating objects are more typical of other disorders, such as Alzheimer's Disease.

# The impact of cognitive and behavioural changes

Cognitive and behavioural changes in MND can significantly impact care and quality of life, including:

**The wellbeing of the person with MND:** People may feel as though they are losing their sense of self and socially withdraw. Behavioural impairments are also associated with depressive symptoms such as negative mood and hopelessness.[19,20]

Although individuals with FTD might be unaware of these changes, apathy still impacts their quality of life.[2] Additionally, FTD, apathy and disinhibition have been linked to reduced survival.[3,21,22]

**The wellbeing of carers and relatives:**[2,5,23–26] Behavioural changes, such as apathy and disinhibition, are strongly associated with carers' perception of loss, anticipatory grief, burden and anxiety.

> **"Carer burden is very high. It will take families 24/7 management to support the person with FTD."**
>
> MND professional

**Service use and treatment adherence:**[20] People with cognitive or behavioural changes may have problems understanding or following care instructions, or understanding why certain therapies might be important. This may lead to issues complying with treatments or using equipment consistently, such as assisted ventilation.

**Decision-making:**[21–23] People with executive dysfunction, attention issues, or mental rigidity may have difficulties making informed decisions. This includes understanding a treatment and the implications of choosing to have, or not have, an intervention.

> **“He can understand simple instructions but gets confused if more than one idea is thrown at him.”**
>
> Carer of a person with MND

**Communication abilities:** Language dysfunction can affect a person’s ability to report a problem, express their needs or communicate their consent to a procedure.[27] They can also affect the ability to use certain communication aids.[28]

**Eating and drinking:** People can develop abnormal eating habits, which could increase the risk of choking for individuals with MND-related swallowing issues (dysphagia).[10]

Cognitive and behavioural changes also affect their ability to follow recommended diets, and cope with a gastrostomy.[29] As a result, the person with MND may be at a higher risk of malnutrition and require more supervision and support to maintain adequate nutrition.

**Health and social care professionals:**[7,23] Cognitive and behavioural changes significantly increase care needs. Executive and behavioural impairments can also negatively impact professionals’ wellbeing and their relationship with the person with MND and their family, increasing the risk of burnout, fatigue and moral distress.[23]

# 2

# Assessing cognitive and behavioural changes

# Identifying changes

The NICE guideline NG42 on MND recommends examining any cognitive and behavioural changes at diagnosis and when concerns arise.[30] Identifying cognitive and behavioural changes early is essential for planning care and helping the person with MND and their family cope with what may lie ahead.[20]

Always involve the person with MND, their family and carers when discussing cognitive and behavioural changes.[30] Be aware that these symptoms may be unexpected in MND, and that people may feel anxious or resistant to discussing them.[24,25]

While the person with MND may not recognise changes in their behaviour, family and friends can often provide valuable insights. However, they may find it difficult to raise concerns privately, as this can feel like a betrayal. Normalising behavioural symptoms as part of MND is therefore essential.

Observing the person's behaviour is also important. When speaking with the person with MND and those close to them, look out for signs such as:

- difficulties managing tasks that were previously easy, for example managing finances
- issues learning something new or finding words in a conversation
- carers saying that the person is not "who they were before", their relationship has changed, they seem distant, uncaring or selfish[24,25]
- carers or family members noticing that the person has become very inflexible or stubborn
- signs of hallucinations or delusions, such as firmly believing something that is not real and becoming fixated on it[24,25]
- difficulties understanding instructions or treatment options when you are explaining an intervention

— disinterest or apathy when you are talking about a procedure that concerns them.

If you notice any of these signs, refer the person for a formal assessment as soon as possible, in line with NICE guidelines on MND (NG42) and dementia (NG97).[30,31]

Assessments are essential to providing support to manage daily living and minimising confusion and frustration. When severe cognitive or behavioural changes occur, formal assessments should be prioritised, as there could be adult and child safeguarding issues.

As some people may not expect non-motor symptoms, it's important to explain these symptoms before any tests. This can reassure the person with MND and those close to them.[3]

> **"It's easier to manage a caring role when you acknowledge that changes to thinking are part of the disease. Once you get your head around this, you find ways to cope and place structure into each day."**
>
> Carer of a person with MND

# Assessment methods

Ideally, all individuals showing signs of cognitive or behavioural changes or FTD should be referred to a neuropsychologist.[32] Where this is not possible, another professional should perform the assessment alone or with a colleague and interpret the results with the help of a neuropsychologist.[6] In this case, the multidisciplinary team should discuss how to conduct, supervise and interpret assessments.

Assessments should cover the symptoms currently defined in the diagnostic criteria for MNDci, MNDbi and MNDFTD (see Section 1). As cognitive and behavioural changes can change over time, regular reassessment should be considered.

Methods to identify possible cognitive and behavioural changes, and whether there is the need for an assessment, include:

- interviews with the person with MND and/or separately with someone who knows them well (informant-based interviews)
- observations and assessment of the person's ability to undertake everyday activities
- standardised screening tools for cognition and behaviour – as other symptoms may mask cognitive and behavioural impairments, screening all individuals can be helpful. See pages 26–27 for a description of validated screening tools.

Different methods can be combined to obtain a comprehensive picture, and they can be used to inform a formal, detailed neuropsychological assessment. This can include:

- interviews
- cognitive tests – where the person completes a series of tasks to assess their cognitive abilities, including memory, executive function, language and visuoperception. For example, tests may assess planning, inhibiting responses and word finding

- questionnaires – carers may be asked to rate the person's behaviour and the presence of emotional lability. People with MND might be asked to rate their own mood to gain an accurate picture of the psychological factors involved.

Any type of assessment should always consider the physical restrictions of MND. For example, dysarthria may cause people with MND to talk slowly and affect their performance in standardised verbal fluency tests.[4]

When interpreting the results, professionals should carefully consider any factors that may cause changes in cognition and behaviour or affect the assessment (See pages 28–33).

> **"You have to take a step back and have a holistic overview of not only the cognitive or behavioural changes, but also how they impact the person's functions and life."**
>
> MND professional

## Screening tools

The table on the next page describes validated tools to screen for cognitive changes, behavioural symptoms or both. They have been designed to minimise demands on speech and motor abilities, which are often impaired in people with MND.[32]

Any health and social care professional can use these tools in the clinic or at home. However, supervision by a neuropsychologist and formal training are recommended.

Keep in mind that various factors may affect a person's test performance (see page 28). If the score falls below the cut-off, the person should be referred for a neuropsychological assessment.

You can access the screening tools described in the table via these links:

- **Edinburgh Cognitive and Behavioural ALS Screen (ECAS):** Training is available through the website and is certified by the European Network for the Cure of ALS (ENCALS) – **ecas.psy.ed.ac.uk/**
- **ALS Cognitive Behavioural Screen (ALS-CBS):** It's available for free in the supplementary materials section on **tandfonline.com/doi/full/10.3109/17482961003727954?scroll=top&needAccess=true**
- **MiND-B:** **ativar.co.uk/en/outcome_measures**
- **ALSFTD-Q:** **eprovide.mapi-trust.org/instruments/amyotrophic-lateral-sclerosis-frontotemporal-dementia-questionnaire**

> **"Screenings should be used as a starting point. A score is not enough. You must look for hints and use the tests together with your clinical observations."**
>
> MND professional

| Tool | What it assesses | Who completes it | Time required | Scoring method<br>NB: scores are indicative | Other notes |
|---|---|---|---|---|---|
| ECAS | — executive function, language, verbal fluency, theory of mind, visuospatial domains, memory[32]<br>— behavioural symptoms typical of bvFTD (via a separate carer interview).[6] | — person with MND<br>— carer. | 20 minutes (but it may take more) | **MNDci:** score ≤ 105/136 (ECAS total score, including memory and visuospatial domains).[6]<br>**MNDbi:** score ≥ 1 in the apathy domain or ≥ 1 in 3/5 domains. [6,33]<br>**MNDFTD:** score ≥ 1 in 3/5 domains.[6,33]<br>NB: these scores refer to the English version. | — It has high sensitivity for detecting mild impairment, FTD and Alzheimer's disease.[32,34]<br>— It can also be used as part of a neuropsychological assessment.<br>— There are alternative versions, including in different languages. |
| ALS-CBS | — cognitive functions<br>— behavioural symptoms (via an informant questionnaire). | — person with MND<br>— informant (carer/ relative/ close friend). | four questions for the person with MND and 15 for the carer (quicker to administer than the ECAS) | **MNDci:** score ≤ 16/20.[6]<br>**MNDbi:** score ≤ 36/45.[33]<br>**MNDFTD:** score≤ 32/45.[6,33] | — People can respond either verbally or in writing, including through speech output devices or eye movements.[6,32] |
| MiND-B | — apathy<br>— disinhibition<br>— stereotypical behaviours. | — carer/ relative/ close friend. | five minutes (nine questions) | **MNDbi:** score ≤ 33/36.[33] | — It's easy to administer.<br>— It can be repeated multiple times. |
| ALSFTD-Q | — behavioural symptoms[33]<br>— MNDFTD. | — carer/ relative/ clinician. | 25 questions, 10 minutes | **MNDbi:** score ≥ 22/100.[33]<br>**MNDFTD:** score ≥ 29/100.[33] | |

# The challenges of assessing cognition and behaviour

Assessing cognitive and behavioural changes can be challenging for different reasons.

**Stigma:** People may feel stigmatised by cognitive impairment and the way it affects their ability to perform previous roles. Some professionals may also think that screening for behavioural or cognitive changes may not be important, or that it may increase anxiety.[4] Normalising cognitive and behavioural changes is essential to provide good care.

**Cultural norms:** They can shape how people behave and how certain symptoms (for example disinhibition or apathy) are reported by both the person with MND and those around them. Remember to take these into consideration when interpreting a test's results or the person's behaviour.

**The subtle nature of cognitive change:** Many people experience mild changes that can be difficult to identify without formal assessment.

**Perceptions:** People may not be aware of cognitive and behavioural changes. They may also be reluctant to say that certain tasks have become challenging, or feel defensive about the concerns reported by family, friends or colleagues.

**Overlapping symptoms:** Other psychiatric or physical symptoms may mask or overlap with cognitive and behavioural symptoms, making diagnoses challenging.[6] See pages 30–33.

> **"People, especially when young or in their working age, might compensate for a lot of lost functions."**
>
> MND professional

Additionally, some symptoms may make it difficult to complete the assessment, such as attention issues or motor and speech impairments. Anxiety about being tested can also affect the assessment score.

Tests can be adjusted as necessary. For example, tools such as the ECAS and the ALS-CBS can be completed either verbally or in writing.

> **"When a person can't complete a formal pen-and-paper questionnaire, I observe them doing a task, such as preparing a meal. I then move on to test more complex functions. For example, I observe them as they process and organise information from a shopping list to find groceries in the shop."**
>
> MND professional

**Language barriers:** Where possible, use interpreters and culturally adapted tools. Both the ECAS and the ALS-CBS tool have been translated into various languages.

**Age and education:** People with limited education and/or reading or writing difficulties (such as dyslexia) may perform less well in certain tests. Consider educational background when analysing test results, especially cognitive assessments.

**Resources (including time, staff and location):** Many teams do not have access to a psychologist/neuropsychologist who can complete a detailed cognitive assessment. This particularly affects people who are only seen in clinics and have subtle cognitive changes.

Remember that cognitive functions and behaviour can change over time, so regular reassessment should be considered.

Tests of behavioural symptoms, such as the MiND-B, can be repeated as they are not completed by the person with MND.

However, tools screening for cognitive changes may be affected by practice. Parallel versions of the ECAS are available to avoid the effects of practising the test multiple times. It is recommended to wait at least 4–6 months in between repetitions.[35]

## What else could it be?

Sometimes, it's difficult to understand if cognitive or behavioural symptoms are caused by brain changes. It's important to assess and rule out anything that can affect someone's behaviour and abilities to think, focus or function.

> **"There are various factors that could cause changes in thinking or behaviour. So you must first start from what it could not be causing them."**
>
> MND professional

**Emotional lability:** Also known as pseudobulbar affect, it causes uncontrollable laughter or crying at inappropriate times.[28,36] These reactions are involuntary and do not reflect what the person feels.

For example, a person might respond inappropriately or laugh at inappropriate times, such as when they are in pain or during a funeral. These reactions can be embarrassing and upsetting, and they might seem callous or unfeeling. They can also be misinterpreted as behavioural inhibition typical of MNDbi or as depression.[6,36]

Emotional lability can affect up to half of people with MND and can occur with or without cognitive changes.[28,36] It's essential to explain that emotional lability can be part of MND. Understanding this symptom can help them manage its impact.

> **“It doesn’t help when people say ‘Get control of yourself’. The more I try to do that, the harder it gets. It feels as though something is using my body against my will.”**
>
> Person with MND

Professionals should discuss potential treatments if the individual feels that emotional lability is causing a problem. Tricyclic antidepressants or selective serotonin reuptake inhibitors (SSRIs) may help.[36] However, they are not always effective and may have unwanted side effects.

**Respiratory issues:** People with MND may experience respiratory problems, such as inadequate ventilation and elevated blood carbon dioxide ($CO_2$) levels. This can lead to confusion, concentration problems, impaired memory and, sometimes, hallucinations.[30] Sleep disturbances (which may be a sign of respiratory problems) can also cause these issues.

Even though these problems are caused by respiratory weakness and not by brain alterations, they could still worsen other cognitive and behavioural issues.

If there are signs of respiratory muscle weakness, respiratory function should be assessed immediately to determine whether non-invasive ventilation might help.[30] If the person already uses a ventilator, make sure it works properly. Check:

- the ventilator’s settings and seals
- whether there are any leaks from the mask, tubes or ventilator’s connections
- whether the mask fits the person correctly, as they might have lost weight
- that the person is using the equipment – cognitive or behavioural changes may affect compliance with treatment.

If the person has severe cognitive problems that may be related to respiratory issues, professionals should:[30]

- ensure that $SpO_2$ is measured (at rest and breathing room air)
- not perform other respiratory function tests (FVC, VC, SNIP and MIP) if the interfaces are not suitable for the person (see page 55).

**Infections:** People with MND may develop respiratory infections, such as pneumonia, or urinary and skin infections. These can cause high fever, pain or difficulty breathing. They may also lead to confusion and issues with reasoning.

**Mood:** Being diagnosed and living with MND is very challenging and can be overwhelming. Emotional distress can be common, with some studies reporting that up to a quarter of people with MND experience depression.[37,38] This can undermine quality of life, increase the risk of social withdrawal and reduce survival.[39–42]

Additionally, low mood can also mask or overlap with behavioural symptoms such as apathy or cognitive ones. For example, it can impact motivation, concentration, and worsen executive dysfunction.[5]

When assessing behavioural changes, it is important to also screen for depression. For example, someone who appears apathetic may score low on MiND-B, suggesting few behavioural symptoms, but may show signs of depression in other assessments.

Considering the two tests' results together, alongside other clinical observations, can help distinguish whether apathy is linked to depression rather than behavioural changes associated with MND.

> **"It's essential to distinguish depression from apathy. Depression comes with sadness and unwillingness to live, while apathy refers to a lack of initiative."**
>
> MND professional

There are various tools to assess depression, including the Hospital Anxiety and Depression Scale (HADS) and the PHQ-9. However, these tools include measurements of physical symptoms like fatigue, which are common in MND and may affect the results.[40,43]

Consider adapting the tool or using the PHQ-2. This is a brief, two-question screening tool that any professional can use to identify signs of depression and refer the person for further assessment.[44,45] Shorter tools like this can also reduce patient fatigue during visits.

**Psychiatric and neurodevelopmental conditions:** Some cognitive and behavioural symptoms seen in MND-bi, MND-ci, or MNDFTD also occur in other neurodevelopmental or psychiatric conditions. For example, psychotic symptoms such as hallucinations are core features of schizophrenia.[46]

Autistic individuals may show stereotypical behaviours, mental rigidity or difficulties with social cognition.[46] Similarly, people with attention-deficit/hyperactivity disorder (ADHD) or dyslexia may experience challenges with executive functions, including verbal fluency, attention, and planning.[47,48]

# Assessing mental capacity[49–52]

Cognitive changes and frontotemporal dementia can impact a person's ability to make decisions. Therefore, it might sometimes be necessary to assess a person's mental capacity.

Assessments must be carried out in line with the Mental Capacity Act (2005) (MCA). This is applicable in England and Wales and empowers anyone to make decisions about their own care and treatment.[49] If you are based somewhere else, refer to the relevant legislation in your area.

The MCA (2005) has five core principles:

- A person must be treated as having decision-making capacity unless it is proven otherwise.
- All possible practical steps must be taken to support and empower a person to make their own decision before concluding they lack capacity.
- A person is not to be treated as lacking capacity merely because they make what others consider to be an unwise decision.
- Any actions or decisions taken on behalf of someone who lacks mental capacity must be in their best interests.
- When making a decision on behalf of someone lacking capacity, it must be considered which decision option will lead to outcomes that are less restrictive of their rights and freedoms.

Assessments can be challenging and time-consuming. Communication difficulties make them more complex as people require more time and individualised support. In this section, we will look at how to assess mental capacity in line with the MCA (2005) and as recommended by the NICE guideline NG108 on decision-making and mental capacity.

To learn more, consider consulting the Code of Practice: **gov.uk/government/publications/mental-capacity-act-code-of-practice**

These resources can also help as they explain mental capacity law in lay terms: **mentalcapacitylawandpolicy.org.uk/shedinars/**

## Carrying out assessment[53]

**When to assess capacity:** Mental capacity should be assessed if there is concern that the person has difficulties making a particular decision. For example, if the person seems unable to understand their options while an intervention is being discussed.

It is important to choose an appropriate time to carry out a capacity assessment, as some people may find this distressing. If the decision is not urgent, the assessment should be delayed until the person is calmer, more responsive, or able to decide independently, where possible. For complex decisions, the assessment may need to be carried out in stages, at a pace that supports the person's ability to understand the relevant information.

**Who should assess capacity:** Mental capacity should be assessed by a trained professional who is familiar with the person, their condition, and the decision.

It can be helpful to involve other staff members who have useful specialist knowledge, for instance the surgeon who will operate on the person (if the decision involves surgery), or a clinician who knows the person well. This also offers the opportunity to reflect together on the outcomes and consider relevant alternative viewpoints.

**How to assess capacity:** While it is important to consider the Code of Practice, case law has expanded our understanding of the approach to assessing mental capacity.[54,55] According to this, the assessment is carried out in three stages (see the flowchart on pages 38–39).

The first stage consists of determining whether the person is unable to make a decision. Then you should determine if there's an impairment in the functioning of the mind or brain. Cognitive changes associated with MND may constitute such an impairment. Note that impairments may also be temporary. For example, alcohol or medications may cause confusion.

Finally, assess whether the person is unable to make a decision because of the identified impairment. Before making a judgment, consider the following:

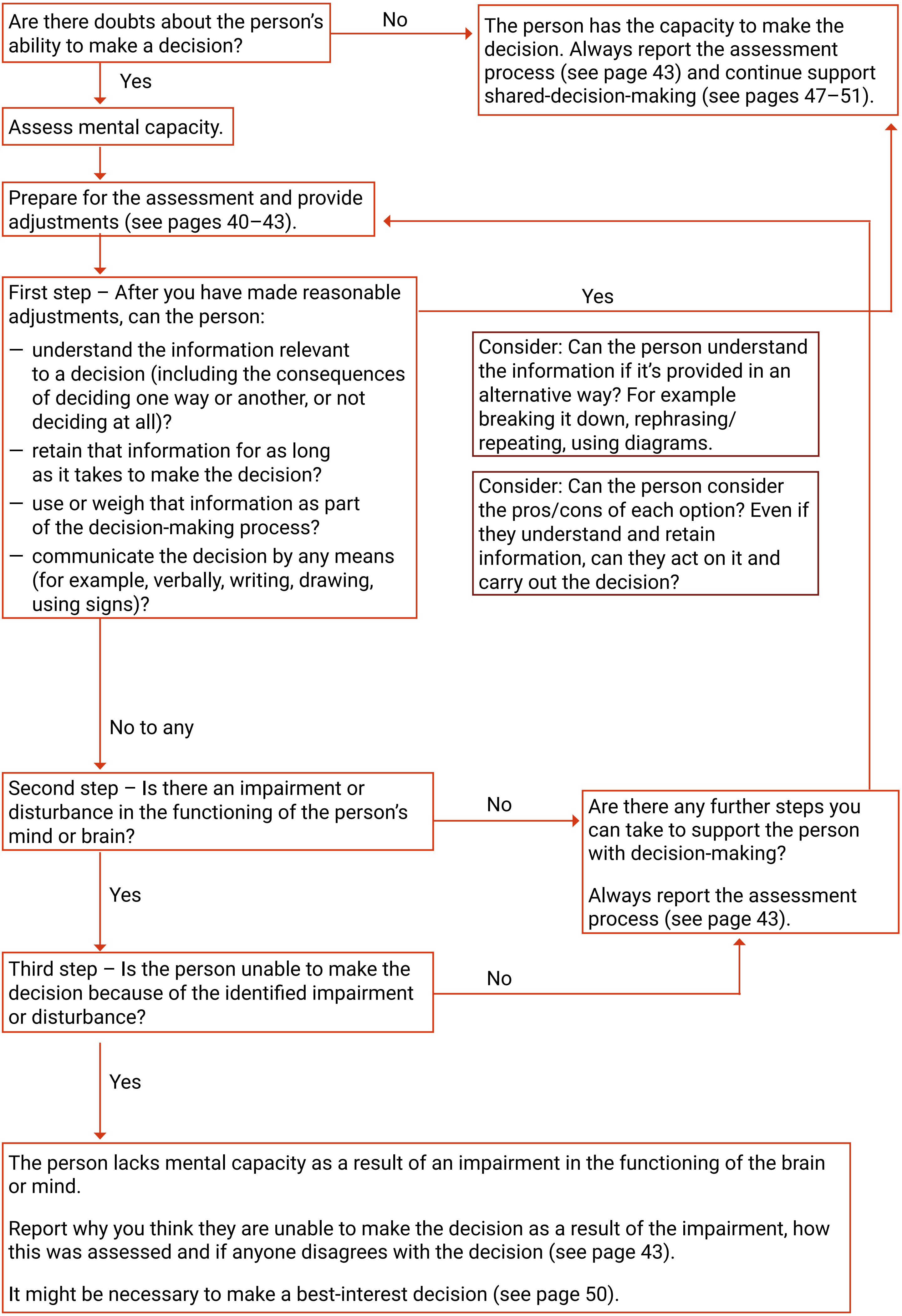
- An assessment only applies to one decision, because capacity is both time and decision-specific. If someone is found to lack capacity, it does not mean they cannot make other decisions, or the same one at a later time.
- A person may have capacity even if they lack insight into their condition.
- Information must be given in a way that is appropriate to the person's circumstances. Reasonable adjustments and support must be provided.
- If the person retains information only for a short period of time, it does not automatically mean they lack capacity. They only need to retain information relevant to the decision and for as long as it's necessary to make the decision.
- If a person makes a decision others consider unwise, it does not mean they lack mental capacity (see page 48–49).
- Take into consideration the person's decision-making history, including the extent to which the person felt involved and listened.

> **“Guide people through the process in a sequential way. For example, if you are discussing gastrostomy, make sure the person understands what it means to come to the hospital and get anaesthesia, or loosing weight and strength if they say no to the intervention.”**
>
> MND professional

It’s essential to take a structured, person-centred, empowering, and proportionate approach to assessment. For this purpose, the assessment process should be adjusted to the person’s circumstances and needs. See pages 40–43.

It may be helpful to follow a checklist for the assessment, such as the MCAST support tool: **bit.ly/MCAST-support-tool**.

The assessment can also be supplemented by real-world observations of the person’s functioning and decision-making abilities, especially if the person has executive dysfunction.



Are there doubts about the person's ability to make a decision?

The person has the capacity to make the decision. Always report the assessment process (see page 43) and continue support shared-decision-making (see pages 47–51).

Assess mental capacity.

Prepare for the assessment and provide adjustments (see pages 40–43).

First step – After you have made reasonable adjustments, can the person:

- understand the information relevant to a decision (including the consequences of deciding one way or another, or not deciding at all)?
- retain that information for as long as it takes to make the decision?
- use or weigh that information as part of the decision-making process?
- communicate the decision by any means (for example, verbally, writing, drawing, using signs)?

Consider: Can the person understand the information if it's provided in an alternative way? For example breaking it down, rephrasing/repeating, using diagrams.

Consider: Can the person consider the pros/cons of each option? Even if they understand and retain information, can they act on it and carry out the decision?

Second step – Is there an impairment or disturbance in the functioning of the person's mind or brain?

Are there any further steps you can take to support the person with decision-making?

Always report the assessment process (see page 43).

Third step – Is the person unable to make the decision because of the identified impairment or disturbance?

The person lacks mental capacity as a result of an impairment in the functioning of the brain or mind.

Report why you think they are unable to make the decision as a result of the impairment, how this was assessed and if anyone disagrees with the decision (see page 43).

It might be necessary to make a best-interest decision (see page 50).

## Preparing and adjusting the assessment

Before starting the assessment, consider:

- Do you have all the necessary information on the decision and the person?
- Does the person understand that their capacity is being assessed?
- Is this the right time and location to carry out the assessment?
- What are the person's needs?
- Have you arranged any necessary adjustments?
- Do you need support from another professional?

The following steps can help you prepare for the assessment.

**Gather relevant information:** Make sure you have all the necessary information on the decision, including the risks and benefits of each decision option, as well as information about the person.
This includes their medical status, care organisation and needs.

- Consult medical notes to find out if the person has any specific support needs.
- Talk with other clinicians involved in the person's care.
- Have informal chats with the person to gain an idea of their needs and thoughts. These conversations can also help you establish a rapport with them, making them feel less anxious.
- Use other tools, such as the ECAS, to identify executive dysfunction, language issues or specific behavioural problems. These should not replace the formal mental capacity assessment.
- Talk with or read reports from carers, family members, care workers, friends or other advocates (with the person's consent).

Sometimes, the person may want a relative or their carer to assist them during the assessment. Ensure that they understand their role is not to "help pass the test" or influence the decisions.

**Provide clear information:** Explain to the person with MND and those around them that capacity is being assessed and why.

Provide information on the decision to be made, the options available and the consequences of deciding one way or another, or not deciding.

Only relevant, salient information should be provided, using clear and accessible language and the person's preferred means of communication. Inclusive communication methods may include:

- gestures or facial expressions
- pointing to objects or people
- using images, videos, drawings or a communication chart
- writing and breaking down information.

> **"Our focus should be on assisting a person to make a decision, not testing their knowledge about it."**
>
> MND professional

**Adjust the assessment:** Make any adjustments to reduce stress, encourage participation, and support decision-making. These could include:

- providing aids, such as glasses, hearing and communication aids
- allocating more time for the assessment
- arranging for emotional support (whether with a professional, carer or relative if the person with MND wants them with them)
- choosing a location where the person is comfortable
- changing the time of the assessment, for example if the person is more alert in the morning rather than the afternoon.

> **“Assessing communication and cognitive abilities is important to understand how the person can be supported to make a decision, and how we should ask questions and present the information.”**
>
> MND professional

**If there are communication issues:** People with MND may experience communication issues due to cognitive changes and/or dysarthria.

Before an assessment, find out if the person uses any communication aids and has a preferred method (such as alphabet board, pen and paper, speech-to-text devices). Some people may also want someone to help them communicate, for example a relative.

Involving a speech and language therapist can be helpful. They can advise on techniques, provide aids and assist you and the person with MND during the conversation.

Consider also that the assessment may take longer. Choose a well-lit, quiet place so that you can both see and hear each other well. This will help you use verbal and nonverbal information (such as facial expressions) to get your messages across. Try to find somewhere private with no background noise. Learn more on: **bit.ly/Supporting-people-with-communication-issues**.

> **“Some issues may be related to communication difficulties, rather than capacity. We must stop and ask ourselves: ‘Have we made this assessment as accessible as possible?’.”**
>
> MND professional

## Recording the assessment

All assessments and decisions should always be documented, regardless of the outcomes. The assessor should record:

- all the steps that were taken to support the person during the assessment
- the outcomes of the assessment and any differences of opinion
- the reasons why a person is considered able to make the decision, and what support they may need to do so. The assessor should work with the person with MND to agree what support they need
- the reasons why a person is considered to be unable to make the decision and how this relates to the identified impairment of the functioning of the brain or the mind
- if they believe that the person's insight or lack of insight is relevant to the mental capacity assessment. They must clearly record what they mean by "lack of insight" in this context and how they believe it affects or does not affect the person's capacity.[56]

See pages 47–51 to learn more about supporting decision-making and best-interest decisions.

# 3

# A practical toolkit

# Working as a multidisciplinary team

Cognitive or behavioural change may not initially impact daily life, but issues may arise when the person faces new challenges or as MND progresses and requires new interventions. Management should focus on planning ahead, considering the likely progression of cognitive and behavioural changes.

Professionals must collaborate to ensure that cognitive and behavioural symptoms are considered in all aspects of care.[7] Working as a multidisciplinary team (MDT) can facilitate communication and collaboration between services.[29,30] If any changes are detected, the whole MDT should be alerted and review the person's care plan. Together, the MDT should:

- raise awareness of cognitive and behavioural changes in MND and signpost the person and their family to relevant support.[30] Our MND Connect Helpline can support (see page 68)
- assess the impact of cognitive and behavioural changes to provide tailored management strategies.[30] Consider that these changes may have a more profound impact if someone suddenly becomes less active. For example, if someone has stopped working after their diagnosis, coping might be particularly challenging and referral the neuropsychologist might be needed earlier
- assess whether the household can manage the changes and arrange support for families and carers.[30] Agree management strategies with the person with MND and their family, and consider that extra checks may be necessary at home to ensure safety
- regularly review any management strategies as cognitive and behavioural symptoms change[30]
- refer to appropriate services. GPs, palliative care teams, community mental health teams, old-age psychiatry, or young-onset dementia services can advise on home support.

- work together to ensure the person can access NHS continuing healthcare (CHC), if they are eligible. Our dedicated webpage can support professionals in this process: **mndassociation.org/chc**.

> **“Justifying cognitive changes within the CHC process can be challenging. Collaborating with different professionals helps gather renewed evidence of the person’s needs.”**
>
> MND professional

Cognitive and behavioural changes can make it challenging to make care decisions, learn how to use new equipment, or adopt routines. It may also affect relationships with family, carers and clinicians. For these reasons, consider the following:

- Advance care planning should be discussed sooner than usual (see page 51).[31,56]
- Interventions such as non-invasive ventilation, communication aids and gastrostomies should be discussed early, potentially earlier than usual, taking into account the progression of both MND and cognitive and behavioural symptoms (see page 54).[30]
- All discussions should be tailored to the person’s needs, considering their communication ability, cognitive status and mental capacity. Additional support may be needed to ensure the person understands the information provided.[56]
- Contingency care plans should be in place if the relationship between the person and their carer breaks down (see page 52).

> **“Management should be preventive rather than reactive. It’s difficult to work once the cognition has deteriorated, so introducing interventions early is very important.”**
>
> MND professional

# Supporting decision-making[56]

Involving people with MND in decisions about their treatment is essential to deliver high-quality, person-centred care. Some individuals need support to make a decision or may lack the mental capacity to do so (see page 34 for how to assess this).

There are various factors that may affect decision-making, including:

- mental health, physical condition and communication needs
- cultural, ethnic and religious factors
- situational, social and relational factors
- the effect of prescribed drugs or other substances.

All members of the MDT should support the person to make an informed decision, adopting a personalised approach.

- Find out how the person wants to be supported and if they want to involve anyone in the decision, and provide reasonable adjustments (see pages 40–42).
- Provide clear, concise and accessible information in Plain English.
- Provide emotional support and ensure the person can express their needs and feel listened to, especially as making certain decisions can be overwhelming and upsetting.
- Be aware of the possible influence and coercion from family members, clinicians or friends. Ensure any help, opinions or explanations are given in a balanced, non-pressuring way.

> **"It's important to consider if the required standards of consent are met. That is, if consent is voluntary and not due to pressure, and if it is informed."**
>
> MND professional

Any decision-making process should be documented and the records shared with the person with MND. Professionals should ask for consent before sharing the records with others.

We offer several resources that can help people with MND make decisions about care: **mndassociation.org/decision-making**.

We also offer resources in various accessible formats and languages, which may help you explain certain interventions (see page 71).

## Supporting people with decisions which seem unwise

People living with MND may sometimes make decisions that others consider unwise. For example, some people may choose to continue eating their favourite foods despite the risk of choking, because those foods are important to their quality of life.

This does not mean they lack mental capacity, and their decision must be respected, even if it carries risks.

> **"As an MDT, we might think a decision is unwise, but it might be totally reasonable from the point of view of the person in our care."**
>
> MND professional

The MDT should ensure the person understands the risks of their decision. They should work collaboratively with the person to reduce risks where possible, including by developing or updating care plans.

The Royal College of Speech and Language Therapists provides guidance on managing situations where a person chooses to have certain foods and drinks despite the risks: **rcslt.org/members/clinical-guidance/eating-and-drinking-with-acknowledged-risks-risk-feeding/**.

Sometimes the carer and family may not agree with the person's decision. The MDT should help them understand the decision and work together to develop an appropriate care plan, if the person with MND agrees.

Discussions with carers and family, and the use of care assessment tools, can help identify whether there is the need for respite care, professional support, specialist referrals or financial assistance.

In some cases, carers or family members may feel unable to support a decision, for example preparing food that carries a choking risk. They have the right to decline, and professionals should ensure that alternative plans are in place to support both the person with MND and their carer.

> **"Relatives are often the ones who are looking ahead. It's important to recognise that the decision of the person with MND may lead to an increase in care needs and be proactive about addressing those needs."**
>
> MND professional

## Making best-interest decisions

When a person lacks mental capacity, professionals can make a decision in the person's best interests. Decisions should centre the individual's wishes, preferences, and values, and involve anyone who can advocate for them.

This could include an attorney appointed by the person and their carer. Be aware that family members and carers may feel making decisions on behalf of the person with MND as a burden, and may need support. Wherever possible, the person with MND should be involved in the decision making process.

Decision-makers should demonstrate that they have determined and prioritised the individual's best interests. All decisions should be clearly documented, including how the person's wishes, preferences and values were considered, and who was involved in the process.

Health and social care services should have systems in place to record a person's wishes regarding interventions, beliefs and values. Advance care planning can help with this.

> **"Best interests should be considered holistically. Someone's faith or meaningful activities need to be given equivalent weight to what we consider to be medical best interests. It is essential to work as a team with whoever can inform such decisions."**
>
> MND professional

## Advance care planning (ACP)

Advance care planning should be discussed early and reviewed regularly with people with MND, ideally before capacity is affected. These conversations can feel overwhelming, particularly around diagnosis, but they help people consider their preferences in advance and maintain a sense of control.

An advance care plan or advance statement can record any information that the individual considers important for their future needs, such as where they wish to receive care. These documents should guide decision-makers if the person later looses capacity. While not legally binding, recorded wishes should be carefully considered and any decision that goes against them should be justified.

Some people may wish to discuss lasting power of attorney. In this case, it's important to discuss which decisions they would like the attorney to make. They may also want to complete an advance decision to refuse treatment. This is a legally binding form that allows the person to specify which treatments they do not want and under what circumstances. Further information is available in our guide "Advance care planning for MND" (see page 71).

# Supporting people close to someone with MND

Supporting carers and family members of someone with MND is essential, particularly when the person experiences severe cognitive or behavioural changes.

> **"You can't change the behaviour. The only thing you can do is support the family to understand, because they can get really traumatised by these changes."**
>
> MND professional

Many carers feel unprepared for these changes. Not understanding why the person with MND is acting out of character can lead to confusion, anger, and sadness. Behavioural changes may also make socialising harder, leaving carers feeling isolated.[24–26] Additionally, carers might misinterpret these behaviours as intentional rather than symptoms, which can be upsetting.[24,25]

Professionals should help carers understand that certain behaviours are symptoms of MND and ensure they feel heard. This can relieve anxiety and prepare them for potential future symptoms, helping them feel empowered and better accept these changes.[24,25] Our information resources can help you with this (see page 71).

Professionals should also assess the needs of carers and family members, including children. Monitor wellbeing with regular check-ins, refer to respite care and mental health support when needed, and offer practical strategies for daily care (see page 56).[20]

Our guide "Supporting children and young people close to someone with MND" can help you assess the needs of children (see page 71).

> **“Exhaustion can creep up on you when you least expect it and tasks that would normally be easy or simple can become very fraught.”**
>
> MND professional

We also offer age-appropriate resources that could help you explain to children why the behaviour of the person with MND changed. Find out more: **mndassociation.org/children-and-young-people**.

Carers may be reluctant to accept outside help, fearing others will not understand the needs of the person with MND. A clear, detailed care plan can provide consistency and reassurance. If external respite is not suitable, it may be helpful to involve other family members, friends, or agencies for home support.

Professionals must also consider risks when behavioural changes include aggression, as this could lead to unsafe situations for carers, family, or the individual with MND. Additionally, behavioural changes can strain or break down the relationship between the person with MND and their carer. Such situations can be challenging for professionals and carers and require careful day-to-day planning and contingency measures.

# Managing MND symptoms

Review our publications listed on the next page to learn more about managing MND symptoms in people with cognitive and behavioural changes or FTD. See also the NICE guideline on MND NG42 on: **nice.org.uk/guidance/ng42**.

**Saliva problems:** If a person with MND has cognitive impairment and saliva problems, consider glycopyrronium bromide as a first-line treatment. This has fewer central nervous system side effects.[30]

**Eating, drinking and swallowing issues:** Pay particular attention to the nutritional and hydration needs of people with MND who have FTD and who lack mental capacity.[30] They may have difficulties following medical advice and may need extra help preparing and eating their meals.

For this reason, the MDT should assess the ability of the person with MND to understand the risks of swallowing difficulties and what support they need from carers.[30]

Enteral feeding options such as gastrostomy should be discussed early and regularly. Conversations should cover the risks of having a late gastrostomy, the benefits of having it fitted early, and the process of refusing or withdrawing from the treatment.[30]

However, be aware that gastrostomies may not be suitable for people with severe cognitive decline.[28] Before deciding on gastrostomy, a neurologist, together with the MDT, should assess the severity of FTD and cognitive issues, the person's ability to make decisions and give consent, and whether they are likely to accept and cope with treatment.[30]

They should also discuss it with the carer and family (with the person's consent, if possible) to ensure there is appropriate support to manage the intervention.[30]

**Respiratory problems:**[30] When assessing respiratory symptoms in people with MND who have FTD, choose the appropriate tests considering the specific needs and circumstances of the individual. These include:

- their ability to give consent
- their understanding of the tests
- their tolerance of the tests and willingness to undertake them
- the impact on family and carers
- whether the person is capable of receiving non-invasive ventilation (NIV).

Before deciding on NIV, the MDT, together with respiratory ventilation services, should assess:

- the person's capacity to make decisions and give consent
- whether the person is likely to accept treatment
- whether the person is likely to achieve improvements in sleep-related symptoms and/or behaviour.

The intervention should also be discussed with carers and families, with the consent of the person with MND (if they are able to give it).

**Further reading:**

Infographic – Respiratory tests for people with MND
Infographic – Dietetics and nutrition
Information sheet P3 – Managing saliva problems
Information sheet P6 – Evaluation and management of respiratory symptoms
Information sheet P8 – Dysphagia in MND

**See page 71 to order publications**

# Practical strategies

This section includes advice on managing cognitive and behavioural changes, with strategies you could suggest to family and carers. It was developed with the help of MND expert professionals.

> **"Management strategies should be individualised and aimed at maintaining function."**
>
> MND professional

## If someone has problems with decision-making and processing complex information:

— Support them and encourage their participation (See page 47).

— Break complex information down into smaller chunks.

— Take time to ensure that the person fully understands each step.

— Check that they understand the consequences of each action or decision.

— If possible, limit the choice to one or two alternatives and avoid open questions. Try asking questions that require yes/no answers.

— If the person is resistant, try phrasing things in different ways.[24,25]

## If someone has problems adhering to treatment:[20]

— Check that the person can understand the instructions.

— Provide clear and accessible written information that may be easier to follow.

— Provide appropriate training for carers and care workers to help them supervise the person with MND.

— Regular in-person, online or phone check-ins and reminders in between visits can help.

### If someone has difficulty learning a new task:

- Encourage them to stop and think.
- Break the task down into smaller steps.
- Use verbal and non-verbal prompts to refocus attention or show them what to do.
- Consider introducing new equipment and communication aids earlier.
- Avoid introducing high-tech communication aids or complex equipment.

> **"Introducing equipment before cognition declines can help the person become familiar with it. This can make it easier to use later, when learning becomes harder."**
>
> MND professional

### If the person has issues with problem-solving:

- Refocus their attention on relevant issues.
- Help them monitor their own performance and provide feedback.
- Try modelling the behaviour you are trying to encourage. For example, demonstrate the task.
- Encourage plenty of practice to reinforce the steps required.

### If impulsivity is a problem:

- Organisation aids (calendars, diaries or reminders) may be helpful.
- Limiting access to credit cards may help if excessive spending or gambling are an issue.
- Supervise activities. People may make decisions too quickly without being careful. Encourage them to stop and think. Visual reminders could help with this.

> **“A person may mistake the front door for the garden door and go out on a busy road. A reminder like a poster saying ‘Use the other door’ can prevent them from impulsively going out.”**
>
> MND professional

### If a person has difficulties finding words:

- Encourage non-verbal responses, such as pointing.
- Consider language issues when recommending communication aids. For example, if someone has issues with spelling, they might not be able to effectively use certain aids. Equipment should be chosen or adapted to accommodate these problems.

### If there are changes in eating habits, suggest the following:[29]

- Supervise the person’s eating more closely to make sure they do not put too much food in their mouth at one time.
- Regularly remind people with MND about tips for swallowing safely (following advice from a speech and language therapist)
- Limit the amount of food on the plate at one time and add food at an appropriate pace to avoid cramming.
- Remove or hide sweets or other foods that the person may impulsively eat.
- Monitor food cravings and involve a dietitian, if necessary.
- Ensure mealtimes are protected from distractions to make sure the person finishes their meal, eats, and drinks enough.

### If the person experiences confusion:

- Establish a structure, such as a timetable or a routine, to help them know what is going on or what to expect.
- Reduce the turnover of carers or staff, including care workers, so that the person becomes familiar with them.

- Reduce noise, stimulation and cluttering.
- Arrange supervision to ensure the person's safety and wellbeing, especially if they live alone.

### If the person is passive and withdrawn:

- Initiate activities and bring the person in.
- Use visual or verbal cues to prompt activities because people may have difficulties initiating a task.
- Establish a structured routine.
- Make sure that the person is not neglecting themselves, especially if they are living alone, and arrange support when necessary.

### If perseveration is a problem:

- Help the person shift their focus to a new task.
- Encourage a calm, structured and orderly environment to help transition to another activity.
- Explain the problem to the carer or family by saying, for example: "Mrs X has difficulty shifting her attention away from an activity once she has started. She will continue to do the same activity even though it is no longer appropriate to the situation. She may appear to be stubborn or not listen properly, but this is due to a problem in her thinking".

### If there are difficulties engaging with everyday tasks:[57,58]

- Focus on one activity at a time.
- Choose the right moment and simplify tasks where possible. If the person with MND is unreceptive, consider delaying the task or skipping it if it causes distress (for example skip shaving).
- Show the person what to do or do it with them, for instance brushing your teeth while they brush theirs.
- Reduce distractions, interference or other stimulation, such as noise.

- Engage the person to help them pay attention and use verbal and non-verbal prompts to refocus attention. Using words such as “just” and “quick” (for example “I just need to put your jumper on, or you’ll be cold”) can make the person with MND feel less overwhelmed by the task and reduce resistance. Explaining the benefits of the activity can also help.
- Break down tasks into separate small steps. Encouraging the person in between steps and using inclusive language when introducing the next step may reduce resistance. For example, try say “Now we are going to brush our teeth”, “That’s it”, “Well done”. Always be mindful of personal space and privacy. For example, standing near enough to help if required, while not standing over the person, can avoid making them feel rushed.

> **“Avoiding behavioural triggers or distractions can help get the person to engage in activities such as getting dressed, being transferred to the wheelchair, going out. Printing a timetable so that they know what’s going to happen could also help avoid outbursts.”**
>
> MND professional

### If the person with MND becomes agitated or aggressive:[57,58]

- Try to understand what triggered that behaviour, such as tiredness or a change in the routine.
- Remain calm and try to distract them. It’s helpful to know the person with MND and their likes and dislikes to avoid triggers or find helpful distractions.
- Avoid getting into an argument.
- Seek support if necessary, such as from another relative or member of staff.

**If there are behavioural problems, such as responding inappropriately or making disinhibited comments:**

- Explain to the carer and family why the person acts inappropriately or seems selfish and uncaring. Reassure them that these reactions or responses are not personal.
- Advise the carer and family that they should express their feelings verbally and as explicitly and simply as possible. Explain that the individual with MND may have problems understanding other people's emotions, thoughts or facial expressions.
- Refer the carer and the family to psychological and emotional support. Helplines and support groups could also be beneficial.
- Suggest removing problematic stimuli that trigger inappropriate behaviours, and refocusing or redirecting the person's attention.
- Arrange for extra support, including respite care and help from psychiatric services if behaviours are challenging.
- Explain to the carers that it might be better to avoid conflict and arguing back. Changing the subject or redirecting the focus might be helpful.[24,25] Walking away for a few minutes may help diffuse the situation.
- Letting friends and family know about personality changes before visits can ease the stress on carers at social events.[24,25]
- Explain that the person's face may become less expressive. If the individual is apathetic, it may be more challenging to understand their feelings or needs. In this case, it is important to observe and report more intuitive signs of distress, such as body positioning, unusual or new behaviour, movement, or non-verbal sounds.

> **"Families will often not understand that this is coming as a result of changes in the brain and not because he has just decided to behave inappropriately."**
>
> MND professional

# References

1. Opie-Martin S, et al. ‘Motor Neuron Disease register for England, Wales and Northern Ireland-an analysis of incidence in England.’ Amyotrophic Lateral Sclerosis & Frontotemporal Degeneration. 2021;22(1–2):86–93. doi:10.1080/21678421.2020.1812661.

2. Radakovic R, et al. ‘Quality of life, cognitive and behavioural impairment in people with motor neuron disease: a systematic review.’ Quality of Life Research. 2024;33(6):1469–1480. doi:10.1007/s11136-024-03611-5.

3. Crockford C, et al. ‘ALS-specific cognitive and behavior changes associated with advancing disease stage in ALS.’ Neurology. 2018;91(15):e1370–e1380. doi:10.1212/WNL.0000000000006317.

4. Abrahams S. ‘Neuropsychological impairment in amyotrophic lateral sclerosis–frontotemporal spectrum disorder.’ Nature Reviews Neurology. 2023;19(11):655–667. doi:10.1038/s41582-023-00878-z.

5. Benbrika S,et al. ‘Cognitive, emotional and psychological manifestations in Amyotrophic Lateral Sclerosis at baseline and overtime: a review.’ Frontiers in Neuroscience. 2019;13:951. doi:10.3389/fnins.2019.00951.

6. Strong MJ, et al. ‘Amyotrophic lateral sclerosis – frontotemporal spectrum disorder (ALS-FTSD): Revised diagnostic criteria.’ Amyotrophic Lateral Sclerosis & Frontotemporal Degeneration. 2017;18(3–4):153–174. doi:10.1080/21678421.2016.1267768.

7. Grand JH, et al. ‘Clinical features and multidisciplinary approaches to dementia care.’ Journal of Multidisciplinary Healthcare. 2011;4:125–147. doi:10.2147/JMDH.S17773.

8. Saxon JA, et al. ‘Examining the language and behavioural profile in FTD and ALS-FTD.’ Journal of Neurology, Neurosurgery, and Psychiatry. 2017;88(8):675–680. doi:10.1136/jnnp-2017-315667.

9. Trojsi F, et al. ‘Theory of mind and its neuropsychological and quality of life correlates in the early stage of Amyotrophic Lateral Sclerosis.’ Frontiers in Psychology. 2016 **frontiersin.org/journals/psychology/articles/10.3389/fpsyg.2016.01934/full**.

10. Turner MR, Talbot K. ‘Sweet food preference in amyotrophic lateral sclerosis.’ Practical Neurology. 2017;17(2):128–129. doi:10.1136/practneurol-2016-001554.

11. Waldö ML, et al. ‘Psychotic symptoms in frontotemporal dementia: a diagnostic dilemma?’ International Psychogeriatrics. 2015;27(4):531–539. doi:10.1017/S1041610214002580.

12. Devenney EM, et al. ‘Unravelling psychosis in motor neurone disease – a study of clinical features, cognition, and survival.’ Journal of Neurology, Neurosurgery & Psychiatry. 2019;90(e7):A6–A6. doi:10.1136/jnnp-2019-anzan.15.

13. Consonni M, et al. ‘Cognitive and behavioural impairment in amyotrophic lateral sclerosis: A landmark of the disease? A mini review of longitudinal studies.’ Neuroscience Letters. 2021;754:135898. doi:10.1016/j.neulet.2021.135898.

14. Lulé DE, et al. ‘Story of the ALS-FTD continuum retold: rather two distinct entities.’ Journal of Neurology, Neurosurgery, and Psychiatry. 2019;90(5):586–589. doi:10.1136/jnnp-2018-318800.

15. Van Langenhove T, et al. ‘Predicting development of Amyotrophic Lateral Sclerosis in Frontotemporal Dementia.’ Journal of Alzheimer’s disease: JAD. 2017;58(1):163–170. doi:10.3233/JAD-161272.

16. Saxon JA, et al. ‘Cognition and behaviour in frontotemporal dementia with and without amyotrophic lateral sclerosis.’ Journal of Neurology, Neurosurgery, and Psychiatry. 2020;91(12):1304–1311. doi:10.1136/jnnp-2020-323969.

17. Gregory JM, et al. ‘Executive, language and fluency dysfunction are markers of localised TDP-43 cerebral pathology in non-demented ALS.’ Journal of Neurology, Neurosurgery, and Psychiatry. 2020;91(2):149–157. doi:10.1136/jnnp-2019-320807.

18. Henstridge CM, et al. ‘Synapse loss in the prefrontal cortex is associated with cognitive decline in amyotrophic lateral sclerosis.’ Acta Neuropathologica. 2018;135(2):213–226. doi:10.1007/s00401-017-1797-4.

19. Rabkin J, et al. ‘Cognitive impairment, behavioral impairment, depression, and wish to die in an ALS cohort.’ Neurology. 2016;87(13):1320–1328. doi:10.1212/WNL.0000000000003035.

20. Caga J, et al. ‘The impact of cognitive andbBehavioral symptoms on ALS patients and their caregivers.’ Frontiers in Neurology. 2019;10:192. doi:10.3389/fneur.2019.00192.

21. Govaarts R, et al. ‘The frontotemporal syndrome of ALS is associated with poor survival.’ Journal of Neurology. 2016;263(12):2476–2483. doi:10.1007/s00415-016-8290-1.

22. Hu WT, et al. ‘Behavior matters — cognitive predictors of survival in Amyotrophic Lateral Sclerosis.’ PLOS ONE. 2013;8(2):e57584. doi:10.1371/journal.pone.0057584.

23. Olesen LK, et al. ‘Reflections of family caregivers and health professionals on the everyday challenges of caring for persons with amyotrophic lateral sclerosis and cognitive impairments: a qualitative study.’ Palliative Care and Social Practice. 2022;16:26323524221077702. doi:10.1177/26323524221077702.

24. Mioshi E, et al. ‘An online intervention for carers to manage behavioral symptoms in motor neuron disease (MiNDToolkit): a randomized parallel multi-center feasibility trial.’ Amyotrophic Lateral Sclerosis & Frontotemporal Degeneration. 2024;25(5–6):506–516. doi:10.1080/21678421.2024.2350658.

25. Mioshi E, et al. ‘“A lightbulb moment”: carers’ experiences of behavioural symptoms in motor neurone disease before and after MiNDToolkit.’ BMC neurology. 2024;24(1):238. doi:10.1186/s12883-024-03746-5.

26. Trucco AP, et al. ‘Factors affecting anticipatory grief of family carers supporting people living with Motor Neurone disease: the impact of disease symptomatology.’ Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration. 2024;25(7–8):776–784. doi:10.1080/21678421.2024.2359559.

27. Paynter C, et al. ‘Communication and cognitive impairments and health care decision making in MND: A narrative review.’ Journal of Evaluation in Clinical Practice. 2019;25(6):1182–1192. doi:10.1111/jep.13219.

28. Hobson EV, McDermott CJ. ‘Supportive and symptomatic management of amyotrophic lateral sclerosis.’ Nature Reviews. Neurology. 2016;12(9):526–538. doi:10.1038/nrneurol.2016.111.

29. Egan A, et al. ‘Dysphagia and mealtime difficulties in dementia: Speech and language therapists’ practices and perspectives.’ International Journal of Language & Communication Disorders. 2020;55(5):777–792. doi:10.1111/1460-6984.12563.

30. Overview | Motor neurone disease: assessment and management | Guidance | NICE. 2016 Feb 24 **nice.org.uk/guidance/ng42.**

31. Recommendations | Dementia: assessment, management and support for people living with dementia and their carers | Guidance | NICE. 2018 Jun 2 **nice.org.uk/guidance/ng97**.

32. Simon N, Goldstein LH. ‘Screening for cognitive and behavioral change in amyotrophic lateral sclerosis/motor neuron disease: a systematic review of validated screening methods.’ Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration. 2019;20(1–2):1–11. doi:10.1080/21678421.2018.1530264.

33. Didcote L, et al. ‘The reliability and validity of in-person and remote behavioural screening tools for people with amyotrophic lateral sclerosis.’ Journal of the Neurological Sciences. 2024;466:123282. doi:10.1016/j.jns.2024.123282.

34. Saxon JA, et al. ‘The Edinburgh Cognitive and Behavioral ALS Screen (ECAS) in frontotemporal dementia.’ Amyotrophic Lateral Sclerosis & Frontotemporal Degeneration. 2020;21(7–8):606–613. doi:10.1080/21678421.2020.1797090.

35. ECAS Downloads – ECAS. **ecas.psy.ed.ac.uk/ecas-downloads/**.

36. Finegan E, et al. ‘Pathological Crying and Laughing in Motor Neuron Disease: Pathobiology, Screening, Intervention.’ Frontiers in Neurology vol. 10 260. 2019, doi:10.3389/fneur.2019.00260

37. Young C,A et al. ‘Prevalence of depression in amyotrophic lateral sclerosis/motor neuron disease: multi-attribute ascertainment and trajectories over 30 months.’ Amyotrophic lateral sclerosis & frontotemporal degeneration vol. 24,1-2 (2023): 82-90. doi:10.1080/21678421.2022.2096410

38. Larsson BJ, et al. ‘Symptoms of anxiety and depression in patients with amyotrophic lateral sclerosis and their relatives during the disease trajectory.’ Journal of the Neurological Sciences. 2023;455:122780. doi:10.1016/j.jns.2023.122780.

39. Woolley SC, Strong MJ. ‘Frontotemporal Dysfunction and Dementia in Amyotrophic Lateral Sclerosis.’ Neurologic Clinics. 2015;33(4):787–805. doi:10.1016/j.ncl.2015.07.011.

40. Gibbons CJ, Young CA. ‘Assessing and Managing Depression and Fatigue in Motor Neuron Disease. Neurodegenerative Disease Management.’ 2012;2(4):401–409. doi:10.2217/nmt.12.42.

41. Bjelica B, et al. ‘Non-motor symptoms in patients with amyotrophic lateral sclerosis: current state and future directions.’ Journal of Neurology. 2024. doi:10.1007/s00415-024-12455-5.

42. Schlüter DK, et al. ‘Risk factors for social withdrawal in amyotrophic lateral sclerosis/motor neurone disease.’ Amyotrophic Lateral Sclerosis & Frontotemporal Degeneration. 2018;19(7–8):591–598. doi:10.1080/21678421.2018.1508477.

43. Faltracco V, et al. ‘Mood disorders in patients with motor neuron disease and frontotemporal symptoms: validation of the Hospital Anxiety and Depression Scale for use in motor neuron disease.’ Journal of the Neurological Sciences. 2025;469:123378. doi:10.1016/j.jns.2024.123378.

44. Garber BB,et al. ‘Using the Patient Health Questionnaire-2 to improve depression screening in head and neck cancer patients.’ American Journal of Otolaryngology. 2023;44(2):103724. doi:10.1016/j.amjoto.2022.103724.

45. Levis B, et al. ‘Accuracy of the PHQ-2 Alone and in Combination With the PHQ-9 for Screening to Detect Major Depression.’ JAMA. 2020;323(22):2290–2300. doi:10.1001/jama.2020.6504.

46. Zucchi E, et al. ‘Psychiatric symptoms in Amyotrophic Lateral Sclerosis: beyond a motor neuron disorder.’ Frontiers in neuroscience vol. 13 175. 2019, doi:10.3389/fnins.2019.0017

47. Farah R, et al. ‘Neuro-behavioral correlates of executive dysfunctions in dyslexia over development from childhood to sdulthood.’ Frontiers in Psychology. 2021**frontiersin.org/journals/psychology/articles/10.3389/fpsyg.2021.708863/full**.

48. Capuozzo A, et al. ‘A systematic review on social cognition in ADHD: the role of language, theory of mind, and executive functions.’ Brain Sciences. 2024. doi:10.3390/brainsci14111117.

49. ‘Mental Capacity Act: making decisions.’ GOV.UK. 2020 **gov.uk/government/collections/mental-capacity-act-making-decisions**.

50. Jayes M, et al. ‘How do health and social care professionals in England and Wales assess mental capacity? A literature review.’ Disability and Rehabilitation. 2020;42(19):2797–2808. doi:10.1080/09638288.2019.1572793.

51. Kim ES, et al. ‘Decision making by people with aphasia: A comparison of linguistic and nonlinguistic measures.’ Journal of Speech, Language, and Hearing Research. 2020;63(6):1845–1860. doi:10.1044/2020_JSLHR-19-00182.

52. Jayes M, et al. ‘An exploration of mental capacity assessment within acute hospital and intermediate care settings in England: a focus group study.’ Disability and Rehabilitation. 2017 Oct 9 **tandfonline.com/doi/abs/10.1080/09638288.2016.1224275**.

53. ‘Decision-making and mental capacity’ | Guidance | NICE. 2018 **nice.org.uk/guidance/ng108**.

54. NHS England. ‘Guidance to support implementation of the Mental Capacity Act in acute trusts for adults with a learning disability.’ **england.nhs.uk/long-read/guidance-to-support-implementation-of-the-mental-capacity-act-in-acute-trusts-for-adults-with-a-learning-disability/**.

55. ‘Discussing the order in which to assess capacity.’ Hill Dickinson. 2022 Dec **hilldickinson.com/insights/articles/discussing-order-which-assess-capacity**.

56. Overview | ‘Shared decision making’ | Guidance | NICE. 2021 **nice.org.uk/guidance/ng197**.

57. Backhouse T, et al. ‘How do family carers and care-home staff manage refusals when assisting a person with advanced dementia with their personal care?’ Dementia. 2022;21(8):2458–2475. doi:10.1177/14713012221123578.

58. Backhouse T, et al. ‘Nurturing attentiveness: a naturalistic observation study of personal care interactions between people with advanced dementia and their caregivers.’ The Gerontologist. 2024;64(6):gnae004. doi:10.1093/geront/gnae004.

# How we can support you

Our MND Connect helpline offers practical and emotional support, information and signposting to people with MND, carers, family and professionals. They can also provide information about our services mentioned below.
**Email:** mndconnect@mndassociation.org
**Tel:** 0808 802 6262

## MND Association website

Our website offers supporting information on MND, our work, services, and how to get involved.
**mndassociation.org/professionals**

Stay updated on events, publications and opportunities for health and social care professionals.
**mndassociation.org/educationupdate**
X: **mndeducation**
Bluesky: **mndeducation.bsky.social**

## Information resources

We produce high quality information for people with MND, carers, families and professionals, available in multiple formats and languages.
**mndassociation.org/pro-info-finder**
**mndassociation.org/careinfofinder**

## Education

Our education programme aims to improve standards of care and quality of life. Opportunities include webinars and face to face equipment training.
**mndassociation.org/education**

## MND Professionals' Community of Practice

A peer led group supporting cross disciplinary learning in MND care. Membership can contribute to CPD and offers access to networking and learning events.
**mndassociation.org/cop**

## Local support

We offer online and local peer support, plus trained volunteers who provide practical help by phone, email or visits.
**mndassociation.org/local-support**

We fund and develop specialist care centres and networks across England, Wales and Northern Ireland, offering multidisciplinary care.
**mndassociation.org/care-centres**

## Financial support

We offer a range of grants to support people living with MND, their families and unpaid carers. These are not in place of any statutory funding that should be available.
**mndassociation.org/getting-support**

## MND register

The Register aims to collect information about everyone with MND in England, Wales and Northern Ireland to support care planning and research.
**mndregister.ac.uk**

## Research into MND

We fund and promote research that leads to new understanding and treatments, and brings us closer to a cure for MND. We also produce information sheets on research for people with or affected by MND.
**mndassociation.org/research**

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# We value your feedback

Your feedback helps improve our information for the benefit of people living with MND and those who care for them. Visit **smartsurvey.co.uk/s/mndprofessionals** or email your comments to **education@mndassociation.org**.

If you would like to help us by reviewing future versions of our information resources, please email us at **education@mndassociation.org**.

# How to order our publications

Our publications are free for anyone with or affected by MND or Kennedy's disease, including professionals. Health and social care professionals can also order items on behalf of someone with or affected by MND or Kennedy's disease.

Download from **mndassociation.org/publications** or contact MND Connect to order hard copies. Call **0808 802 6262** or email **mndconnect@mndassociation.org**.

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**Every day we support people affected
by Motor Neurone Disease.
Because with MND, every day matters.**