

Cognitive change, frontotemporal dementia and MND



Motor neurone disease (MND) is a progressive and terminal disease that results in degeneration of the motor neurones, or nerves, in the brain and spinal cord.¹

MND also includes non-motor symptoms, one of the most common being cognitive change. Research has shown that changes can occur in the frontal and temporal areas of the brain, which affect thinking, reasoning and behaviour.²

This booklet contains information on cognitive and behavioural change and dementia, and practical tips on management. It has been designed to support your work in helping people with MND, families and carers adjust to changes in thinking and behaviour, should they happen.

Information for people affected by MND

- *Will the way I think be affected?* (information sheet for people with MND)
- *How do I support someone if their thinking is affected?* (information sheet for carers of people with MND)
- *Managing emotions* (information sheet for people with or affected by MND).

Download from our website at **www.mndassociation.org/publications** or see page 34 for details of how to order copies.

How the MND Association can support you

The MND Association supports professionals to care for people affected by MND in a variety of ways, which includes providing:

- a range of information and educational opportunities
- local support and advice from our staff and volunteers.

MND Connect

Our helpline offers information and support by telephone and email – on **0808 802 6262** or **mndconnect@mndassociation.org**

See page 34 for more information about how the MND Association can support you in your role.

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Overview of cognition and MND

People with MND appear to fall into one of four groups:³

- around 50% are unaffected by cognitive change
- around 35% experience mild cognitive change, with specific deficits in executive functions, language and/or social cognition^{2,4,5,6}
- up to 15% develop frontotemporal dementia (FTD), either at the same time or after diagnosis of MND⁷
- up to 15% of people diagnosed with FTD go on to develop MND. Symptoms of dementia may lead to FTD being diagnosed before movement is affected and MND is diagnosed.

Note: While figures are based on current evidence, there is some suggestion that the number of people with MND affected by cognitive change may be higher. Numbers are dependent on the sensitivity of the tool used to detect cognitive change.

International criteria on the diagnosis of cognitive change in MND uses the following categories:

- MND (the person has MND but is unaffected by cognitive change or FTD)
- MNDbi or MNDci (MND with behavioural impairment or cognitive impairment)
- MND-FTD (MND and frontotemporal dementia).⁸

There is a wide spectrum of changes in cognition in MND. Some people experience very mild changes, barely noticeable, whereas for others the changes can be more pronounced and obvious.²

Cognition refers to a range of high-level brain functions that can be separated into a number of different areas:

Executive functioning

- Setting and achieving goals.
- Planning and problem solving.
- Responding to new situations.
- Shifting attention or dividing attention between different tasks.
- Initiating and inhibiting responses.

Language

- Being able to understand and produce (in speech or writing) language (the underlying organisation of words).

Memory

- Acquiring, retaining and retrieving new information.

Perception

- Dealing with the information gathered by the five senses.

Social cognition

- Understanding and interpreting other people's thoughts/ beliefs/feelings.

In those with MND affected by cognitive change, the most prominent impairment is in executive functions. These functions are particularly dependent on the frontal lobes of the brain. Some people also show changes in language and behaviour, and some researchers have questioned whether language changes may be at least as common as executive function changes.⁹

Changes in social cognition can be challenging for carers/family of people with MND, as well as for health and social care professionals.

Memory is not usually affected, although some people may experience problems with their memory as a secondary consequence of executive dysfunction, along with problems with attention. Problems with perception are rarely seen.

A small but significant minority of people experience severe changes that will be diagnosed as frontotemporal dementia (FTD).² FTD is an increasingly recognised form of dementia, with different signs and symptoms to the more common Alzheimer's disease.¹⁰ People with MND are not protected from developing other forms of dementia, such as Alzheimer's, but there is a very clear link between MND and FTD. See page 16 for more information on FTD.

Some people will first present with cognitive features, which may include behavioural changes, emotional lability (not related to dementia) or frontotemporal dementia, and then go on to display symptoms of MND. In this group, the dementia may mask physical symptoms, because of the person's difficulty recognising and verbalising that something is wrong; hence the importance of physical examination.

Progression of cognitive change has been investigated by some research studies, although results are mixed.¹¹ Research has shown shorter survival in those with cognitive change, when compared with those with intact cognition.¹²

A recent study has shown that behavioural changes (apathy, disinhibition and stereotypical behaviour) are pervasive and do not affect survival.¹³ These behavioural changes, however, are strongly associated with carer burden.^{14, 15}

Inherited MND (sometimes known as familial MND) accounts for approximately 5-10% of all cases of MND.¹ Mutations in the gene C9orf72 have been shown to be the most common cause of inherited FTD, MND and MND-FTD.^{16, 17} Importantly, up to 10% of cases of sporadic MND (where there is no known family history) are also strongly associated with this same gene.¹

In some cases, genetic testing is available to individuals with a family history of MND and/or FTD.

Information for people affected by MND

- Research information sheet B – *part 1: an introduction to inherited MND*
- Research information sheet B – *part 2: genetic testing and insurance*

Download from our website at **www.mndassociation.org/researchsheets** or see page 34 for details of how to order copies.

Experience and recognition of symptoms from different perspectives

People with MND

Someone experiencing cognitive change may have insight in the early stages that something is wrong. The person may recognise that known tasks they used to complete with ease are now more challenging. They may find it more difficult to organise their activities or finances, or think of new ways to solve problems. They may also be aware of difficulties in finding words and understanding complex sentences. They may or may not be aware that learning new tasks is more difficult or that new information is more difficult to absorb and understand.

Family members, friends and carers

It is likely that carers, family members and friends may realise that something has changed. There may be confusion, however, about the changes they perceive and whether they are a feature of MND or perhaps a reaction to a changing situation. They may feel concerned and protective, and may be worried about broaching the subject of cognitive and/or behavioural change.

The family may experience relief when cognitive and behavioural changes are professionally acknowledged, and the relationship to MND is properly attributed and explained. A brief introduction to cognitive and behavioural changes is important prior to assessment, as patients and their families may not understand why an assessment of memory, for example, is needed.

It is crucial that family members, partners and carers are involved in any assessment of cognitive change, as they will have a perspective on the person's past and present behaviour and personality, and any changes that have occurred. It is important that they are asked to try to identify changes in cognition and behaviour that cannot simply be attributed to the physical symptoms the person is experiencing.

Health and social care professionals

Professionals may notice difficulties when giving instructions or explaining procedures. Apathy and disinterest or ambivalence regarding intervention may give clues.

It may be noticeable that, when discussing important clinical consequences of MND, families demonstrate strong emotional reactions, while the patient seems unconcerned. This may be in contrast to any reaction they demonstrated in initial visits when receiving confirmation of diagnosis.

For health professionals it may be difficult to untangle a mix of symptoms that could be misinterpreted as stubbornness or inflexibility, rather than as signs of underlying cognitive change.

In some cases, symptoms may be picked up by family and professionals, but not properly understood, at a variety of key points in the disease journey. For example, when decisions have to be made about home adaptations, the person's reaction to or understanding of the need for changes may be unexpected. Another example could be when decisions are being made about gastrostomy, and the person cannot understand the complexity of the information given.

If you recognise any of the symptoms detailed over the next few pages, perhaps in combination, it is advisable to discuss them with the person with MND and their carer/family (where relevant and appropriate) to see if they are aware of any changes and to explore their thoughts. It is important to rule out other causes. Any changes should be discussed with the person's neurologist and the wider multidisciplinary team.

Any health or social care professional can make use of tools such as the Edinburgh Cognitive and Behavioural ALS Screen (ECAS), MiND-B or M-ACE (see pages 18-20) to screen for potential signs of cognitive or behaviour change. If the result indicates some evidence of impairment, then advice should be sought from local clinical neuropsychology services.



A person with MND initially refused a gastrostomy, so his family wanted to respect his wishes, despite their own preferences and understanding of the importance of timing. Around this time, the patient was assessed for symptoms of cognitive change and it was confirmed he had MND-FTD. His refusal of gastrostomy was revealed to be related to changes in his ability to reason and discuss things, whereas in the past he would have discussed it with his family and sought more information to make his decision." A clinician

Cognitive and behavioural changes

Cognitive change in MND that is not dementia involves subtle and specific deficits mainly, but not exclusively, in executive function, along with behavioural changes.

Executive dysfunction

Executive function is the ability to set and achieve goals, to review and monitor performance and to adapt according to change and feedback. Deficits in executive function are of similar nature to those seen in frontotemporal dementia (FTD – see page 16) but are milder in severity.

Everyday impact of executive dysfunction

In day-to-day life, a person with executive dysfunction may experience difficulties with:

- multi-tasking
- organising themselves and timekeeping
- making and implementing plans
- setting goals
- concentration and distractibility
- finding solutions to problems and correcting mistakes
- making decisions
- initiating ideas
- sequencing, organising and monitoring performance of tasks
- generating ideas and thinking flexibly
- inhibitive and controlling thoughts.

These can affect the ability to:

- manage affairs/finances
- plan for the future

- concentrate, for example when reading or dealing with household bills
- undertake new activities, use new equipment or learn new tasks (which may have implications on interventions such as communication aids, gastrostomy and assisted ventilation)¹⁸
- hold a conversation if background distractions are present
- do more than one thing at a time, eg ironing while watching television
- manage a sequence of activities
- complete work, leisure and self-care activities
- live alone without support
- adapt to having an illness and make decisions about its management.

These issues can be combined with changes in behaviour and social awareness (see opposite).

Impact on carers

Cognitive change and behavioural symptoms are strongly associated with higher levels of burden and increased stress for people with MND, family members and carers. It is important to assess for the presence of cognitive change as early as possible after symptoms are recognised and to identify ways to help those affected.^{14, 15}

Language dysfunction

Impaired rapid word generation (verbal fluency) is reported in almost all studies of cognitive change in MND.^{19, 20} Although this is often used as a test of executive function, and people may score poorly due to other issues (such as anxiety or depression), impaired verbal fluency can also indicate problems with language that are related to cognitive change.

Verbal fluency deficits are more prominent in those with pseudobulbar palsy, but they are not restricted to people with this bulbar-onset form of MND.²¹ At times it can be difficult to differentiate the changes in language that are due to bulbar deficits, and those that are dependent on cortical brain changes. The changes related to bulbar function may mask, for some time, those related to cognitive change.⁹

The deficits are of a similar nature to those seen in frontotemporal dementia (FTD – see page 16) but are milder in severity. They include:

- reduced verbal expression and initiation (not due to dysarthria)

- impaired naming of objects, including difficulty with finding the name of objects presented to them
- perseveration - repetition of a word or phrase that is no longer appropriate to the situation, and the use of stereotyped expressions
- echolalia - repeating parts of another person's speech that have just been heard
- word-finding difficulty in conversational speech – when people pause to search for an appropriate word or name. This may lead to circumlocution, where people talk around a word as they search for it
- semantic paraphasias – mixing up names for closely associated objects, eg 'spoon' instead of 'fork'
- phonological paraphasias - where people say part of an intended word, eg pun instead of spun
- difficulties understanding complicated sentences
- impaired comprehension of words – sometimes worse for verbs than nouns
- problems with spelling.^{9,22}

Problems with language can affect selection and use of communication aids.

MND-aphasia

Some people may show primary language impairment, in comprehension and/or production, similar to primary progressive aphasia. It may be the presenting feature in some cases and can occur without personality changes.²³

Changes in behaviour and social cognition

Behavioural impairment is a recognised feature of MND.

Behavioural problems may include:

- behavioural disinhibition: socially inappropriate behaviour, disinhibited comments, loss of social manners
- acting impulsively without thinking, inability to delay gratification (may include gambling/inappropriate internet shopping)

- apathy and inertia: being withdrawn and distant, lacking interest, not initiating activities^{24, 25}
- loss of sympathy and empathy for others: reduced response to other people's needs and feelings (including their partner or carer, if they have one), and social cues. Reduced interest in others and social warmth, 'not the same person as before' – see section opposite on Theory of Mind^{26, 27, 28}
- perseverative, rigid, stereotyped or compulsive/ritualistic behaviour: simple repetitive movements, use of stereotypical phrases, uncontrolled repetition of a response (eg a catchphrase) or behaviour
- hyper-orality and dietary change: overeating/cramming, altered food preference (often for sweet foods), excessive drinking or smoking.

Someone may act in a way that is quite different to their previous self. For example, they may make tactless comments to people. Alternatively, they may act in exaggeration of previous traits, for example, changing from being determined to being stubborn and inflexible. They may become restless, irritable and in some cases aggressive.

Note: people who are cognitively normal can nonetheless have profound behavioural abnormalities.

The deficits above are of similar nature to those seen in frontotemporal dementia (FTD) but are milder in severity. At least three of the examples above should be seen to meet the criteria for behavioural variant FTD (bvFTD – see page 16).

Social awareness and loss of insight

Due to loss of insight, the person affected by cognitive change may be less aware of any changes in themselves. Carers or family, however, may be acutely aware of the changes in behaviour. It is not uncommon to hear comments such as, 'They are not the person they were.' Carers or family may also be confused by the changes they are observing and whether they are caused by MND, depression or frustration, or problems with their relationship etc.

For this reason, it is important to rule out other causes that can affect someone's ability to concentrate and function, such as fluctuations in mood, changes in breathing, ineffective use of a ventilator or presence of infection (see 'What else could it be?').

Theory of Mind

Theory of Mind is the ability to infer mental state (thoughts, feelings, desires and intentions) in other people, and to understand that other people think differently and have different mental states. It also relates to judgements based on the behaviour and emotional expression of another person. Impairment in Theory of Mind can be an early sign of cognitive change. Assessment of Theory of Mind (included within the ECAS Tool – see page 18) can be beneficial for early identification of the behavioural form of FTD.^{29, 30, 31}

Memory dysfunction

Memory impairments in MND usually involve immediate recall and problems lie in the 'taking in' of information rather than forgetting. Memory changes may also be noted in frontotemporal dementia, but these are more severe in nature.

Visuospatial function

Perceptual processes are largely preserved in both frontotemporal dementia and milder cognitive change. Some problems have been noted with visual perception, but those with mild cognitive change often have little difficulty finding their way around, with spatial orientation and locating objects.

Neuroanatomical changes and imaging

The brain changes found in cognitive decline in MND are of a similar but more subtle nature than those in FTD. Changes are seen in both structure (grey and white matter) and function (blood flow). These changes are particularly found in the frontotemporal regions and the motor cortex.

Such abnormalities have been related directly to impairments in cognition (verbal fluency and doing two things at once). Findings from MRI scans indicative of atrophy also suggest that this may be a biomarker of cognitive impairment.^{32, 33, 34, 35}

What else could it be?

Mood

The progressive nature of MND presents people with the condition with a continual need for psychological adjustment.

Adapting to physical problems can lead to changes in mood and most people experience frustration, anger and distress. For others, the emotional change can be more profound and result in depression. Changes in engagement with tasks or concentration may be related to low mood rather than cognitive change. There is also evidence that delay in diagnosis can lead to greater risk of depression.³⁶ Management may include counselling and medication.

The occurrence of depression in people with MND is often lower than might be expected, given the nature of the illness.³⁷ This impression may be due to how depression is assessed in people with MND, but its level of occurrence has been shown to be similar to that seen in people with other neuromuscular disorders, and should not be ignored.³⁸

Emotional lability

Some people with MND experience emotional lability (also known as pseudobulbar affect), which can result in uncontrollable laughter or crying in response to something that is only moderately funny or sad (eg a television programme). There may be inappropriate responses at embarrassing times (eg laughing during a funeral), or inability to respond appropriately to other people, which may be interpreted as strange, callous or unfeeling. This can be disturbing to the person themselves and to those around them.

Emotional lability is reported to affect 19-49% of people with MND and can occur in those with or without cognitive change. At present, there is no evidence that lability is associated with cognitive changes.³⁹

It is important to reassure the person with MND and family/carers that this is a factor of the condition. Some people find that understanding the symptom helps them to manage the impact. Others find that emotional lability limits where they go and what they do.

Treatment should be discussed and instigated only if the individual feels that emotional lability is causing a problem. Tricyclic antidepressants or SSRIs (eg fluoxetine) may alleviate this symptom, although this isn't always successful.⁴⁰

Information for people affected by MND

- Information sheet 9C - *Managing emotions* includes more about emotional lability for people affected by MND.

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Respiratory involvement

Weakening of breathing muscles caused by MND leads to inadequate ventilation and a build-up of carbon dioxide (CO₂) in the blood. Hypercapnia (raised CO₂), along with disrupted sleep (another symptom of respiratory muscle weakness) can cause changes in concentration, memory and lead to confused thinking.

Non-invasive ventilation has been shown to improve some of these symptoms.^{41,42} It is important to note that these respiratory-related deficits are not the same as cognitive and behavioural changes in MND, which at present cannot be treated with ventilation or pharmacological approaches.

Assessment of respiratory function is recommended if there are symptoms of respiratory muscle weakness, to determine whether a trial of non-invasive ventilation might be beneficial. If the person is already using a ventilator to help support breathing, it is important to check settings and make sure the ventilator is working properly. Hypoventilation can result from underutilisation of a ventilator, so check settings and seals, look out for leaks in the mask, tubing and connections, and check the person is using the equipment. Those with cognitive or behavioural change may have problems with following instructions and complying with treatment.⁴³

- See our information sheet P6 - *Evaluation and management of respiratory symptoms in MND*.

Download from our website at www.mndassociation.org/publications or see page 34 for details of how to order copies.

Infections

In acute-onset infections (eg chest or urine), the signs of infection, such as high temperature, feeling unwell, etc. may be accompanied by confusion.

Frontotemporal dementia (FTD)

A small minority of people with MND show changes in cognition and behaviour of sufficient severity to warrant a diagnosis of MND-FTD.⁸ These changes are most often in behaviour and executive skills, but sometimes they are in language or conceptual understanding.

There are three main variants of FTD, which are referred to as:

- behavioural variant (bvFTD)
- progressive non-fluent aphasia (PNFA) or non-fluent variant PPA (nfvPPA)
- semantic dementia (SD) or semantic variant PPA (svPPA).

The variants are not mutually exclusive. Some people show a mixture of problems in language and behaviour.

People with MND-FTD will not typically experience all possible symptoms of MND, or all possible symptoms of one of the types of FTD described below.

Instead, they tend to present with a combined set of symptoms that are usually from the bvFTD form, along with other language symptoms that could be related to PNFA or SD.

Behavioural variant FTD (bvFTD)

The principal characteristics are similar to those seen in milder cognitive change (see pages 11-12), but more severe in nature.^{44, 45, 46}

At least three of the examples shown on pages 11-12 are required to meet criteria for bvFTD. Usually, behavioural changes and executive deficits are both present, but they may vary in their relative prominence. Sometimes, people will show severe changes in social behaviour and yet perform relatively well on standard tests of executive function.

Tests of Theory of Mind (included within the ECAS tool – see page 18) can be of value in such cases in detecting early cognitive change.

Note:

- The changes should not be explicable in terms of the physical restrictions caused by MND.
- Behavioural changes such as apathy and inertia and loss of feelings for others should not be secondary consequences of depression, fatigue or respiratory difficulties in MND.

Progressive non-fluent aphasia (PNFA)^{47, 48}

Here, the principal difficulties are in expressive language, although understanding of grammar can also be affected. The main characteristics are:

- reduced generation of language
- impaired use of grammar in language production
- word retrieval difficulties (naming of verbs may be more affected than nouns)²²
- sound-based errors
- impaired understanding of syntactically complex sentences.

Understanding of individual words is well preserved.

Note:

Problems in expressive language (aphasia) need to be distinguished from motor speech difficulties (dysarthria) resulting from the physical changes of MND.

Semantic dementia (SD)^{49, 50}

Within MND, this type of FTD is extremely rare. The problem lies in understanding of concepts. The principal characteristics are:

- problems in naming and understanding words
- semantic errors in naming (eg dog instead of tiger)
- fluent, effortless but empty, circumlocutory speech output
- relative preservation of understanding of syntax
- difficulty in recognising faces and objects.

Semantic problems are associated with atrophy of the temporal lobes, which can be more marked on the left or right side.⁵¹

Assessing cognitive and behavioural change

Timely assessment is important. Understanding the level of cognition of the person with MND is crucial to help them and their family to cope with what may lie ahead, including being able to make timely decisions about treatment and care.

Assessments can lead to suggestions of how to help minimise any confusion and frustration that the changes are presenting. Any management of changes in cognition and/or behaviour should always involve the person with MND, together with their family and carers.⁵²

Severe cognitive and behaviour change may have implications for adult and child protection issues, so assessment should be prioritised. Such assessments can inform (but not replace) assessments of capacity.

Assessment methods

Assessment methods commonly used by professionals include:

- interview
- functional assessment
- standardised screening measure of cognition and behaviour
- informant-based behavioural questionnaires
- assessment according to current criteria for MNDbi (behavioural impairment), MNDci (cognitive impairment) and MND-FTD.⁸

A formal, detailed assessment should be undertaken by a clinical neuropsychologist where available.

ECAS tool

The Edinburgh Cognitive and Behavioural ALS Screen (ECAS) tool has been designed specifically as a first step in assessing the presence of cognitive change in MND.⁵³

The tool is designed to take 20 minutes to complete by any health or social care professional, including non-neuropsychologists. It tests functions that may be impaired in MND, including executive functions, language and fluency, as well as those not usually affected: memory and visuospatial skills. It has been designed to take account of physical disability, as answers can be written or spoken. Part of the assessment includes a carer interview about the behaviour of the person with MND.

Download the ECAS tool from <http://hdl.handle.net/1842/6592> Guidelines for use and a training video are also available on this website. Completion of ECAS training is certified through ENCALS (European Network for the Cure of ALS).

The tool can be used to screen people to see whether they would benefit from a full neuropsychological assessment.

Using the ECAS tool

- The ECAS is a screening tool. If someone falls below the cut-off score, referral should be made for full neuropsychological assessment.
- Where full neuropsychological assessment is not possible or is not suitable for the person with MND, the results of ECAS screening should at least be interpreted with the help of a neuropsychologist.
- Performance on the ECAS by the person being screened may be affected by age and education. People with poor schooling and/or reading or writing difficulties will do less well. This must be taken into consideration in the interpretation.
- How the tool is used and how usage is supervised should be discussed as a multidisciplinary team.
- Beware of the label of cognitive impairment and what it means for the person with MND and their family. It should not affect equity of care.

MiND-B

The MiND-B is a simple tool for the identification and quantification of behavioural symptoms in ALS.⁵⁴ It measures three behavioural domains: apathy, disinhibition and stereotypical behaviour. It consists of nine questions with a total score of 36. Higher scores denote absence of or very mild behavioural symptoms. The MiND-B can be completed by a carer, family member or clinician. It can be downloaded from www.neura.edu.au/forefront/mind

M-ACE

The M-ACE is a brief and sensitive cognitive screening tool for dementia.⁵⁵ Unlike the ECAS tool, it is not specific to MND. It can be downloaded from www.neura.edu.au/frontier/research/test-downloads

Neuropsychological assessment

Assessment methods commonly used by clinical neuropsychologists include:

- interview
- detailed cognitive assessment – this involves a person completing a series of tasks that assess their cognitive abilities. Assessments may include tests of memory, executive functions, language and visuoperception (eg planning, generating and inhibiting responses, understanding sentences and word finding)
- questionnaires – carers may be asked to rate the person's behaviour and the presence of emotional lability. People with MND may be asked to rate their own mood in an attempt to gain an accurate picture of psychological factors involved.

The challenges of assessing cognitive change

Challenges that may delay identification of cognitive changes in MND include:

- the stigma associated with cognitive impairment and the serious implications it has in terms of someone's ability to carry out former roles
- the subtle nature of cognitive change in the majority of people who may be affected, which means it can be difficult to identify within the clinical setting
- a lack of self-awareness and concern about cognitive and behavioural change – a person may not be motivated to report dysfunction and may be defensive about dysfunction reported by family or colleagues.

The challenges in terms of completing the assessment include:

- motor and speech impairments, which often mask cognitive difficulties and render it hard to carry out assessment

- the time, location and resources needed to assess this aspect of functioning (particularly for those people only seen in clinic who have subtle cognitive changes). Many teams do not have access to a clinical psychologist/neuropsychologist who is able to complete a full and detailed assessment of a person's cognitive function
- while diagnosis of cognitive impairment depends on at least two tests of executive function,⁸ assessment should also look at changes in other domains (eg language)
- cognitive function can change over time, so reassessment should be considered.



Listen to carers when monitoring for change.
For example, if a carer says, 'My partner is different, he just doesn't love me anymore' – this may well be a change in the relationship since diagnosis, but it could also indicate something deeper in terms of behaviour and lack of empathy."

A carer whose husband had MND-FTD

Supporting people affected by cognitive change and dementia

Being alert to the possibility of cognitive and/or behavioural change at any stage of MND can be valuable, as these changes may have an impact on service use and decision making.

Management of people who show signs of cognitive impairment should focus on forward planning and organising appropriate support strategies for them and their families/carers.

If cognitive change has been identified, it is important to alert all members of the multidisciplinary team, allowing them to react and implement any changes needed in care planning.

For example:

- discussions around interventions and advance care planning may be started sooner rather than later, with additional support to aid and check understanding
- a speech and language therapist may avoid introducing high-tech communication options
- professionals may choose to give information that is more simple and succinct
- levels of support provided around the home may increase and checks may be required to enable safety in the home – eg removing or locking away items that may be used inappropriately
- clinical neuropsychology services, where available, may be involved in care.

Ongoing reassessment of needs is essential. Cognitive or behavioural change may not affect daily life at first, but issues may emerge when the person faces new challenges.

Professionals who can support management of cognitive change

If the person is not already in contact with them, it may be useful to refer to occupational therapy (for strategies and equipment to manage activities of daily living) or speech and language therapy (for strategies and equipment for communication).

The following services may be referred to for home support:

- GP
- palliative care team
- community mental health team
- old-age psychiatry services
- young-onset dementia services
- local multidisciplinary team.

Advance care planning

Many people living with progressive illnesses such as MND fear losing control and not being able to make their own care decisions. Discussion of advance planning is recommended, particularly if cognitive change is identified.

One of the topics discussed may be advance decision to refuse treatment (ADRT). An ADRT is a decision someone can make in advance to refuse specific treatments in certain circumstances in the future. This can include the right to refuse life sustaining treatment. An ADRT tells people about those decisions and becomes active when the person loses the ability to make decisions. It is up to the person with MND whether they choose to complete an ADRT.

It is important to document any discussions so that the person's wishes are respected, particularly in the absence of any formal statements, such as an ADRT.

Information for people affected by MND

- Information sheet 14A – *Advance Decision to Refuse Treatment (ADRT)*
- *End of life guide*

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Supporting carers and family members

Supporting carers is vital. Their individual needs may be complex depending on the severity of cognitive and/or behavioural change in the person with MND.

Cognitive and behavioural change is a symptom for which many carers feel unprepared, especially as many people will not have been told that this can occur as part of MND. Clear explanations and instruction can help. The needs of carers and family members should be assessed and support strategies advised.

The need for respite may be essential, but may be complicated by the carer's concern for their loved one. They may worry that other people will not understand or interpret the person's needs properly while they, as primary carer, are absent.

A clear and detailed care plan is essential, so the carer feels supported and reassured that they have been listened to and that instructions are consistent and sensitively understood. Calling on family, friends and agencies that can provide support within the home may be more helpful if external respite is felt to be less appropriate.

Professionals must be aware of the risks to carers and family where behavioural changes include aggression. A combination of lack of empathy and self-seeking behaviour may lead to carers and family or the person themselves being in danger, especially if the person has retained mobility. Situations such as these will be challenging for professionals, as well as carers, and day-to-day management must be considered.



Someone always had to be there with my husband, so I had to find help just to be able to make our lives work. We had various care workers to support, but it wasn't always successful ... I'd have to wait until he was asleep – only then could I concentrate on organising things or getting tasks done like ironing. I lived on adrenalin and learnt to leave things that weren't essential, like gardening."

A carer whose husband had MND-FTD

Management strategies

These guidelines, created by Professor Sharon Abrahams, may help identify and manage what can be disturbing changes for patients and carers.

Supporting people with cognitive or behavioural problems

- Remember that difficulty processing information, making decisions and/or behaviour problems may be a result of cognitive change.
- If cognitive change is evident, this may interfere with informed decision-making and learning to use new equipment or new routines.
- Where cognitive change is evident, use simple and straightforward language, with closed rather than open questions in order to communicate clearly and directly.
- Consider whether cognitive change is causing a problem. Look at particular areas, for example work, home or relationships.
- Be aware that some people will have severe cognitive problems, some very mild and many will have no cognitive problems.
- Reduced activity and fewer demands on effective cognitive functioning, for example if the person has stopped working or has increased reliance on others, may mean that cognitive change affects them less profoundly.

Has the person experienced symptoms of respiratory impairment?

Problems with concentration, memory and confused thinking related to respiratory insufficiency (and not to cognitive change) may be improved with a trial of non-invasive ventilation.⁴¹

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

- ensure that decision-making is not taken away, but supported: provide an appropriate level of help with decision-making processes and to ensure informed consent
- break down complex information into smaller chunks
- take time to ensure thorough understanding at each step
- take time to check there is understanding of consequences of each action or decision.

Also: People with MND and frontotemporal dementia may lack mental capacity. Capacity issues should be assessed where FTD is evident and care should be provided in line with the Mental Capacity Act 2005

For simpler decision-making:

- limit choices to one or two alternatives
- do not use open-ended questions. Instead, ask questions that require yes or no answers.

If someone has difficulty learning a new task:

- encourage them to stop and think
- reduce the cognitive load by breaking down the task into small steps
- use verbal/non-verbal prompts to help at each step, or try to refocus attention or show them what to do.

Help to problem-solve by:

- refocusing attention on relevant issues
- helping them to monitor their own performance
- helping to provide feedback
- encouraging plenty of practice to reinforce the steps required.

Also: consider implications for introducing new equipment and communication aids.

If impulsivity is a problem:

- suggest organisation aids, such as calendars, diaries or reminders

- supervise activities. People may make decisions too quickly, without remembering to be careful or to use safety equipment. Encourage them to stop and think.

If there appear to be word-finding difficulties (language impoverishment):

- encourage non-verbal responses, such as pointing
- try modelling the behaviour you are trying to encourage, eg demonstrating the task.

Also: consider the implication of language problems on provision of appropriate communication equipment.

If the person is passive and withdrawn:

- they are likely to have difficulty initiating activities
- use visual or verbal cues to prompt activity
- aim for a structured routine.

If perseveration is a problem:

- help to refocus on a new task
- encourage a calm, structured and orderly environment
- explain the problem to the carer/family in terms such as: '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 listening properly, but this is due to a problem in her thinking'.

If the person experiences difficulty getting ready or organised for the day:

- focus on one activity at a time
- engage interest and remove distractions
- break down tasks into discrete steps
- use verbal and non-verbal prompts to refocus attention or show what to do
- minimise interference.

If there is a noted change in eating habits:

- supervise the person's eating more closely
- people with more severe changes may place too much food in their mouth at one time and cram food, while others may eat more food than they need
- limit the amount of food on the plate at one time
- ask the carer to model eating at an appropriate pace
- if food cravings are noticeable, question how much of a problem the behaviour is causing. It may be helpful to discuss with a dietitian.

If there is a noted change in eating habits caused or compounded by bulbar weakness:

- those with poor swallowing may have trouble following medical advice to modify consistency or to thicken drinks
- refer to speech and language therapy for assessment and advice about how to encourage safe eating, eg using the chin-tuck technique, counting to 10 when swallowing
- repeated reminders about swallow safety tips may be necessary
- ensure that mealtimes are protected from any distractions.

If someone responds inappropriately to carer/family etc:

- those affected by cognitive change may have trouble distinguishing facial expressions
- support the carer/family in understanding the reasons behind what appears to be an inappropriate/uncaring response
- advise the carer/family that they should express their feelings verbally and as explicitly/simply as possible
- recommend that they check understanding and repeat as necessary.

Also: the person's own face may become less expressive. If behaviour is apathetic, there may also be increased problems in recognising what the person is feeling or requires. In this situation, it is important to take note of more intuitive signs of distress, eg body positioning, unusual or new behaviour, movement or non-verbal sounds.

If egocentricity is evident (eg loss of concern for carer/family):

- support carer/family in understanding the reasons behind apparent selfishness and explain that this is a problem in the person's thinking
- explain this is not personal
- encourage extra support and regular respite.

If socially inappropriate behaviour is evident (eg laughing, loss of control, sexually disinhibited comments):

- explain to carer/family that this is part of the disease, to foster understanding and support
- explain that studies have shown reduced understanding of emotion and social situations
- there may be misinterpretation of other people's expressions, for example, difficulty recognising the difference between happy and surprised facial expressions
- there may be difficulty in understanding the emotions and thoughts of others
- support the carer/family with strategies to refocus/redirect attention and to deal with aggressive behaviour
- seek help from psychiatric services if challenging behaviour emerges.

Finally, the needs of children should be kept in mind as there may be a need to explain changed behaviour in age-appropriate terms they can understand.



Even with FTD, my husband could understand what was being said to him and make choices if the conversation focused on one thing at a time. I'd tell professionals not to muddy things by talking about lots of subjects, but just to stick to one."

A carer whose husband had MND-FTD

Summary

All health and social care professionals should grasp the opportunity to empower and educate people with MND on cognitive change and FTD.

Knowing that cognitive change can be part of MND and the disease process may come as a relief to people with MND and carers who have noticed changes in thinking and behaviour since diagnosis, but may have been previously reassured that MND does not affect the mind.

If cognitive change is suspected, it is important to understand how it affects the individual, so that strategies and plans can be put in place to care for them appropriately. This can help the person to feel more in control. It also supports family members and carers to understand what is happening and why, and empowers them to help the person that they care for.

Be aware of how you discuss and describe cognitive change and ensure that someone is not 'labelled' with cognitive change. It should not affect equity of care.

Involvement of professionals across the multidisciplinary team is crucial to ensure broad discussion and awareness from different professional perspectives. Discussion should include the person and their family, so that distinction can be made between normal responses in the face of changes caused by MND, and the subtle yet distinct changes attributable to underlying cognitive change. It also ensures that any suggested strategies are applied consistently and suit individual and family routines and lifestyle.

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Further reading

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More information on the categorisation of cognitive change and dementia in MND can be found in Strong MJ et al. Consensus criteria for the diagnosis of frontotemporal cognitive and behavioural syndromes in amyotrophic lateral sclerosis. *Amyotrophic Lateral Sclerosis*. 2009; 10:131-146.

A cognitive change and frontotemporal dementia reading list is available to download from www.mndassociation.org/professionals or can be ordered from the MND Connect team on **0808 802 6262**.

How the MND Association can support you

We support health and social care professionals to provide the best possible care for people living with MND, their carers and families. We do this in a number of ways:

MND Connect

Accredited by the Helplines Standard, MND Connect offers information and support, and signposting to other services and agencies.

Telephone: **0808 802 6262**

Email: **mndconnect@mndassociation.org**

Information resources

We produce high quality information resources for health and social care professionals who work with people with MND. We also have a wide range of resources for people living with and affected by MND.

Downloads of all our information sheets and most of our publications are available from our website at **www.mndassociation.org/publications** or you can order our publications directly from the MND Connect team.

MND Association website

Access further information at **www.mndassociation.org/professionals**

MND Association membership

Join us now and help fight MND. Membership costs £12 for individuals. It is free for people with MND and for carers, spouses or partners of people living with MND. Call **01604 611855** or email **membership@mndassociation.org**

MND support grants and equipment loan

Where statutory funding or provision has been explored and is not available, we may be able to provide a support grant or equipment loan. Our support grant service consists of providing care and quality of life grants for people with MND. This service is supported by MND Association branch and group funds, and by the Association's central fund.

Our equipment loan service is focused on three core items

- riser-recliner chairs
- specialist communication aids
- portable suction units.

For suction units, a small charge is made to statutory services for carriage, maintenance and cleaning.

Referrals for support grants or equipment loan need to be made by a relevant health or social care professional. Call the Support Services team on **01604 611802**, email **support.services@mndassociation.org** or visit **www.mndassociation.org/getting-support**

Research into MND

We fund and promote research that leads to new understanding and treatment and brings us closer to a cure.

Contact the Research Development team on **01604 611880** or **research@mndassociation.org**. Alternatively, visit **www.mndassociation.org/research**

For the latest research news, visit our research blog at **www.mndresearch.wordpress.com**

Our peer-to-peer research and care community blog (RECCOB) has a number of reporters who write updates on MND-related workshops and events at **www.reccob.wordpress.com**

International Symposium on ALS/MND

Each year we organise the world's largest clinical and biomedical research conference on MND. It is the premier event in the MND research calendar for discussion on the latest advances in research and clinical management. Visit **www.mndassociation.org/symposium**

Local support

Regional care development advisers

Our network of regional care development advisers (RCDAs) have specialist knowledge of the care and management of MND. They work closely with local services and care providers to ensure effective support for people affected by MND, provide education for health and social care professionals in MND, and are champions at influencing care services.

MND care centres and networks

We fund and develop care centres and networks across England, Wales, and Northern Ireland, which offer specialist clinical expertise from diagnosis onwards.

Branches and groups

We have volunteer-led branches and groups nationwide providing local support and practical help to people with MND and their carers.

Association visitors (AVs)

Association visitors are volunteers with experience of MND who provide one-to-one local support to people affected by MND.

Other useful organisations

The Frontotemporal Disease Support Group

This group is particularly directed towards carers who are coping with behavioural changes in a partner, family member or friend as a result of frontotemporal dementia. **www.ftdsg.org**

The familial Frontotemporal Dementia Support Group (fFTDSG)

This group offers information, advice and social opportunities for people affected by inherited forms of FTD. This group holds a national meeting once a year in London. **www.ftdsupport.org**

FTD Talk

Information and factsheets about FTD. **www.ftdtalk.org**

International contacts

The Association for Frontotemporal Degeneration (US)
www.ftd-picks.org

Feedback form

Please send us your feedback on *Cognitive change, frontotemporal dementia and MND*

Thank you for taking the time to provide your feedback on one of our information resources.

This questionnaire can be accessed online if preferred, using the following link: www.surveymonkey.com/s/cognitivechange

What is your profession or specialism?

Did you find this resource useful?

Yes Somewhat Not really No

Please explain your answer

Will this information resource help you to provide people affected by MND with any of the following? (tick all that apply)

- an increased understanding of their symptoms
- an increased understanding of their condition
- more independence
- an increased ability to raise awareness of their needs
- more confidence
- improved quality of life
- a greater ability to maintain dignity

Continued overleaf

Feedback form continued

Were there any particular topics that were useful to you?

Was there any information that you didn't find useful or relevant?

Are there any other MND-related topics you would like more information about?

Would you be happy to help us improve our information by becoming an expert reviewer?

Yes (please include your email address below) No

Do you have any experiences of working with people with MND you could share as an anonymous quote or case study for future resources?

Yes (please include your email address below) No

Please return your completed form to:

Education and information team

MND Association

PO Box 246

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Name:

Email:

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If you'd like to help us by reviewing future versions of this or other resources, please email us on infofeedback@mndassociation.org

About us

The MND Association was founded in 1979 by a group of volunteers with experience of living with or caring for someone with MND. Since then, we have grown significantly, with an ever-increasing community of volunteers, supporters and staff, all sharing the same goal – to support people with MND and everyone who cares for them, both now and in the future.

We are the only national charity in England, Wales and Northern Ireland focused on MND care, research and campaigning.

Our mission

We improve care and support for people with MND, their families and carers.

We fund and promote research that leads to new understanding and treatments, and brings us closer to a cure for MND.

We campaign and raise awareness so the needs of people with MND and everyone who cares for them are recognised and addressed by wider society.

MND Association

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About MND

- MND is a fatal, rapidly progressing disease that affects the brain and spinal cord.
- It attacks the nerves that control movement so muscles no longer work. MND does not usually affect the senses such as sight, sound and feeling.
- It can leave people locked in a failing body, unable to move, talk and eventually breathe.
- It affects people from all communities.
- Some people may experience changes in thinking and behaviour, with a proportion experiencing a rare form of dementia.
- MND kills a third of people within a year and more than half within two years of diagnosis.
- A person's lifetime risk of developing MND is up to 1 in 300.
- Six people per day are diagnosed with MND in the UK.
- MND kills six people per day in the UK.
- It has no cure.

mndconnect
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