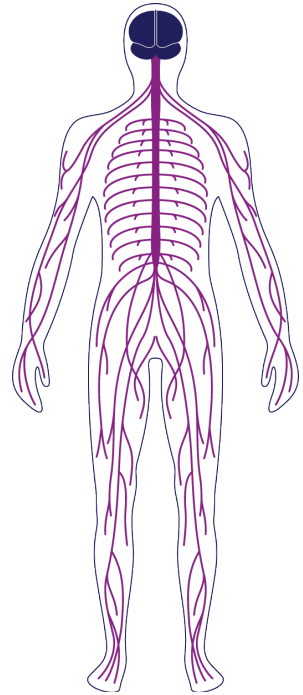




**Cognitive change,  
frontotemporal dementia  
and MND**

# 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.
- It can leave people locked in a failing body, unable to move, talk and breathe.
- It affects people from all communities.
- Some people may experience changes in thinking and behaviour, with some experiencing a rare form of dementia.
- MND kills a third of people within a year and more than half within two years.
- 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.



## Would you like to find out more?

Contact our helpline MND Connect if you have any questions about MND or want more information about anything in this booklet.

**mndconnect**  
**0808 802 6262**  
[mndconnect@mndassociation.org](mailto:mndconnect@mndassociation.org)

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This guide has been endorsed by the MND Professionals' Community of Practice.

# Introduction

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.<sup>1</sup>

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.<sup>2</sup>

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, and their families and carers, adjust to changes in thinking and behaviour, should they happen.

## Information to share

*Changes to thinking and behaviour with MND* booklet

Download from [www.mndassociation.org/publications](http://www.mndassociation.org/publications) or contact MND Connect to order hard copies. Call 0808 802 6262 or email [mndconnect@mndassociation.org](mailto:mndconnect@mndassociation.org)



# Overview of cognition and MND

The general term 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 tasks, initiating and inhibiting responses.

**Language:** being able to understand and produce language (the underlying organisation of words) in speech or writing.

**Behaviour and social cognition:** understanding and interpreting other people's thoughts, beliefs and feelings.

**Memory:** acquiring, retaining and retrieving information.

**Perception:** dealing with the information gathered by the five senses.

See pages 9-13 for more detailed information about how these functions may be affected. In MND, the most prominent impairment is typically in executive functions. These functions are particularly dependent on the frontal lobes of the brain. Some people also show changes in language, social cognition and/or behaviour.

Some researchers have questioned whether language changes may be at least as common as executive function changes.<sup>10</sup>

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

## People with MND appear to fall into one of four groups:<sup>3</sup>

- around 50% are unaffected by cognitive change; this decreases to 20% in the final stage of the disease<sup>4</sup>
- around 35% experience mild cognitive and/or behaviour change, with specific deficits in executive functions, language and/or social cognition.<sup>2, 5, 6, 7</sup> Behaviour changes, including apathy and disinhibition, can occur with or without cognitive changes<sup>8</sup>
- up to 15% develop frontotemporal dementia (FTD), either at the same time or after diagnosis of MND<sup>9</sup>
- up to 15% of people originally diagnosed with FTD go onto to develop MND. In these people, dementia is 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 or behaviour change may be higher. Numbers may be dependent on the sensitivity of the tool used to detect cognitive change and on the stage of the disease.<sup>4</sup>

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

- MND (the person has MND but is unaffected by cognitive or behaviour change or FTD)
- MNDbi (MND with behavioural impairment), MNDci (MND with cognitive impairment) or MNDcbi (a combination of the two)
- MND-FTD (MND and frontotemporal dementia).<sup>8</sup>

There is a wide spectrum of changes in cognition and/or behaviour in MND. Some people experience very mild changes which are barely noticeable, whereas for others the changes can be more pronounced.<sup>2</sup>

A small but significant minority of people experience severe changes that will be diagnosed as frontotemporal dementia (FTD, see page 18).<sup>2</sup>

FTD is an increasingly recognised form of dementia, with different signs and symptoms to the more common Alzheimer's disease.<sup>11</sup> These changes are most often in behaviour and executive skills, but sometimes they are in language or conceptual understanding.<sup>12</sup>

# Experience and recognition 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 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 that they sometimes struggle to find words and understand complex sentences. They may be aware that learning new tasks is more difficult, or that new information is harder to absorb and understand. For those with more severe change and FTD such insight will be affected and these people will not be aware of such changes.

## Family members, friends and carers

Carers, family members and friends may realise that something has changed. They may be uncertain, however, about whether the issues they perceive are a feature of MND, or perhaps a reaction to a changing situation. They may be concerned it is their relationship that is changing. They may also feel protective and 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 is needed. It should be explained that cognitive and/or behaviour changes can occur in some people.<sup>63</sup>

It is crucial that family and carers are involved in any assessment. They will have a perspective on the person's past and present behaviour and personality, and changes that have occurred. Typically, they are interviewed separately from the person with MND. It is important they 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. The NICE Guideline on MND (NG42) highlights the importance of considering cognitive change when considering treatments and therapies, as the person may not understand the complexity of the information given when decisions are being made about interventions such as gastrostomy or assisted ventilation.<sup>38</sup>

Apathy and disinterest or ambivalence regarding intervention may give clues. Cognitive difficulties may manifest as signs of stubbornness or inflexibility, which are misinterpreted by family members and health professionals.

Health or social care professionals should make use of tools such as the Edinburgh Cognitive and Behavioural ALS Screen (ECAS), ALS Cognitive Behavioral Screen (ALS-CBS) or MiND-B (see pages 22-25) to screen for potential signs of cognitive or behaviour change. However, training and/or supervision is advisable before undertaking them.





# Cognitive and behavioural change

Cognitive change in MND that is not dementia involves subtle and specific deficits, mainly in executive and language functions, along with behavioural changes.<sup>12</sup>

## Executive dysfunction

Executive functions include the ability to set and achieve goals, to review and monitor performance and to adapt according to change and feedback. Deficits in executive functions are of similar nature to those seen in FTD (see page 18) but are milder in severity.

In day-to-day life, executive dysfunction may cause difficulties with:

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

This 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 for interventions such as communication aids, gastrostomy and assisted ventilation)<sup>13</sup>
- 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 page 12-13).

## Language dysfunction

At times it can be difficult to differentiate the changes in language which are due to physical bulbar deficits (dysarthria), and those which relate to cognitive change. The changes related to bulbar function may mask, for some time, those related to cognitive change.<sup>10</sup>

Impaired rapid word generation, such as generating names of animals or words beginning with the letter 'A' (verbal fluency) is reported in almost all studies of cognitive change in MND and is assessed in screening tests such as the ECAS.<sup>14, 15</sup> 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 people with MND affected by pseudobulbar palsy, but they are not restricted to people with this symptom.<sup>16</sup>

Language deficits and dysfunction are of a similar nature to those seen in a type of FTD (see page 18) but are milder in severity.

They include:

- reduced verbal expression and initiation (not due to dysarthria)
- recently developed problems with spelling. This can be a prominent and common symptom and will affect whether people with severe dysarthria can use communication aids<sup>10, 17</sup>
- impaired naming of objects, including difficulty with finding the name of objects presented to them

- perseveration – repetition of a word, phrase or action 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
- difficulties understanding complicated sentences
- impaired comprehension of words – sometimes worse for verbs than nouns.

### **MND-aphasia**

Some people may show marked and severe primary language impairment. It may be the presenting feature in a small proportion of cases and can occur without personality changes.<sup>18</sup>



## Changes in behaviour and social cognition

Behavioural impairment is a recognised feature of MND. Research has shown that behavioural changes (apathy, disinhibition and stereotypical behaviour) are pervasive and do not affect survival.<sup>19</sup>

Behavioural problems may include:

- apathy and inertia – being withdrawn and distant, lacking interest, not initiating activities<sup>20, 21</sup>
- 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/buying from cold callers)
- 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'<sup>22, 23, 24</sup> (see section below on Theory of Mind)
- perseverative, rigid, stereotyped or compulsive/ritualistic behaviour – simple repetitive movements, use of stereotypical phrases, uncontrolled repetition of a response (eg a catchphrase) or behaviour, checking or hoarding
- 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, previous traits may become exaggerated, for example, changing from being determined to being stubborn and inflexible. They may become restless, irritable and in some cases aggressive. This combination of symptoms may appear to be a personality change.

Apathy is often the most prevalent of behaviour changes and manifests as a loss of interest in social situations, activities and daily life. It is particularly related to problems in initiating thoughts and actions.<sup>64</sup>

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

It may be unclear whether changes in cognition or behaviour are the direct result of brain changes in MND. It is important to rule out secondary 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?' on page 15).

## **Theory of Mind**

Social cognitive abilities are all the thinking processes we use to interact with others. Theory of Mind is the ability to infer the mental state (thoughts, feelings, desires and intentions) of 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 pages 22-24) can be beneficial for early identification of the behavioural form of FTD.<sup>25, 26, 27</sup>

## **Other changes**

### **Memory dysfunction**

Memory impairments in MND usually involve recall, and problems lie in the 'taking in' and/or retrieving of information rather than forgetting.

### **Visuospatial function**

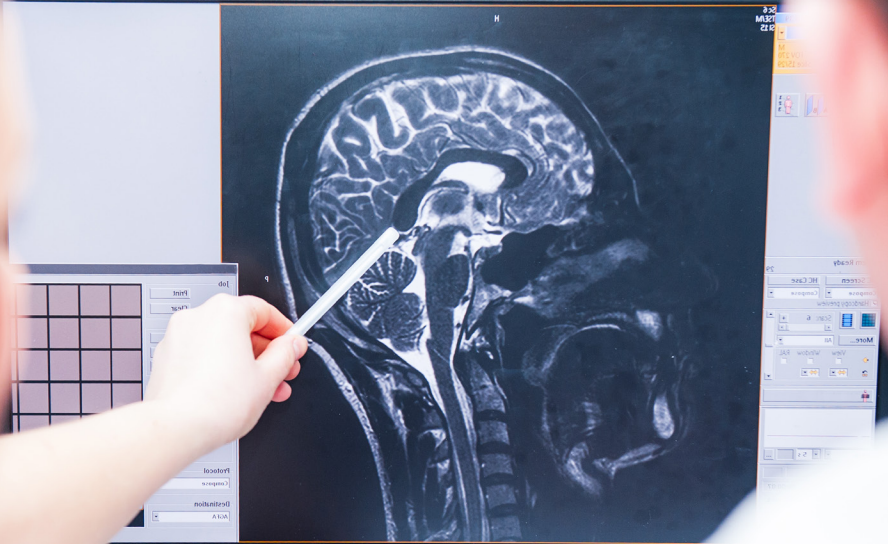
Perceptual processes are largely preserved in both MND and FTD. Problems with spatial orientation and locating objects are more typical of other disorders, namely Alzheimer's Disease.

## 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.<sup>28, 29, 30, 31</sup>

Cognitive changes as found using tools such as the ECAS have been shown to be related to pathological changes in specific parts of the brain. These include problems with a build-up of an abnormal protein which is found in both MND and FTD<sup>65</sup> and problems with the connections between nerve cells in the brain or synapses based on post-mortem pathology studies.<sup>32</sup>



# What else could it be?

## Mood

The progressive nature of MND presents people 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.<sup>33</sup> Management may include counselling and/or medication.

The occurrence of depression in people with MND is often lower than might be expected, given the nature of the illness.<sup>34</sup> This 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 neurodegenerative conditions, and should not be ignored.<sup>35</sup>

## Emotional lability

Emotional lability (also known as pseudobulbar affect) refers to the uncontrollable expression of emotion, such as uncontrollable laughter or crying, which may not reflect what the person is feeling inside.

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 both to the person 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.<sup>36</sup> It is important to reassure the person with MND and family/carers that this is an accepted feature of the condition. Some people find that understanding the symptom helps them to manage the impact.

Treatment should be discussed and instigated only if the individual feels that emotional lability is causing a problem. Tricyclic antidepressants or selective serotonin reuptake inhibitors (SSRIs - eg fluoxetine) may alleviate this symptom, although this isn't always successful and may have unwanted side effects.<sup>37</sup>

### Information to share

*Emotional and psychological support* booklet

Download from [www.mndassociation.org/publications](http://www.mndassociation.org/publications) or contact MND Connect to order hard copies. Call 0808 802 6262 or email [mndconnect@mndassociation.org](mailto:mndconnect@mndassociation.org)

## 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.

## Respiratory involvement

Weakening of breathing muscles caused by MND leads to inadequate ventilation and a build-up of carbon dioxide (CO<sub>2</sub>) in the blood. Hypercapnia (raised CO<sub>2</sub>), 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.<sup>38, 39</sup> It is important to note that these respiratory-related deficits are not the same as cognitive and behavioural changes in MND, which are related to changes in the brain. However, respiration problems may exaggerate symptoms.

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.<sup>40</sup>

### **Information for you**

Information sheet P6 - *Evaluation and management of respiratory symptoms in MND*

### **Information to share**

Information sheet 8A – *Support for breathing problems*

Information 8B – *Ventilation for MND*

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# Frontotemporal dementia (FTD)

People with MND are not protected from developing other forms of dementia, but there is a very clear link between MND and FTD. Some people will first present with cognitive or behaviour features, or FTD, 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.

The question arises whether people with MND may develop FTD with progression of disease even when they show no or only mild cognitive changes early on. Progression of cognitive change has been investigated by some research studies, although results are mixed. Cognitive and behaviour change can occur at the beginning of the disease but recent research shows that they are even more common in later stages of the disease.<sup>4,41</sup> Research has shown shorter survival in those with cognitive change, when compared with those with intact cognition.<sup>42</sup>

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

- behavioural variant (bvFTD), the most typical presentation in MND<sup>43</sup>
- progressive non-fluent aphasia (PNFA) or non-fluent variant primary progressive aphasia (nfvPPA)
- semantic dementia (SD) or semantic variant PPA (svPPA).

Patients may also present with a combined set of symptoms that are usually in the form of bvFTD, along with other language symptoms.

## Behavioural variant FTD (bvFTD)

A diagnosis of MND-FTD requires evidence of progression of symptoms in cognition and/or behaviour. In addition at least three of the examples of behaviour change shown on page 12 are required to meet the criteria.

Alternatively, at least two of those cognitive/behavioural symptoms need to be identified, along with a loss of insight and / or psychotic symptoms.

An additional means of diagnosing MND-FTD is to identify the presence of language impairment meeting criteria for either progressive non-fluent aphasia (PNFA) or semantic dementia (SD) (see headings below) and this may occur alongside symptoms of cognitive/behavioural change<sup>8</sup>

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. In such cases, tests of emotion recognition can be of value 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)<sup>44, 45</sup>**

Here, understanding of individual words is well preserved. 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)<sup>18</sup>
- sound-based errors
- impaired understanding of syntactically complex sentences.

**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)<sup>46, 47</sup>

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.<sup>48</sup>

Symptoms of psychosis are generally rare in FTD and therefore not included as part of clinical diagnostic criteria. Nevertheless, these symptoms can occur in some people with MND, MND-FTD and FTD, particularly those who have a mutation in the C9ORF72 gene.<sup>49, 50</sup>

## Genetic testing

Inherited MND (sometimes known as familial MND) accounts for approximately 5-10% of all cases of MND.<sup>1</sup> Mutations in the gene C9ORF72 have been shown to be the most common cause of inherited FTD, MND and MND-FTD.<sup>51, 52</sup> Importantly, up to 10% of cases of sporadic MND (where there is no known family history) are also strongly associated with this same gene.<sup>1</sup>

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

### Information to share

Research information sheet B1 - Inherited MND: introduction

Research information sheet B2 - Inherited MND: genetic testing and insurance

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# Assessing cognitive 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.

The NICE guideline on MND<sup>38</sup> recommends that a person's multidisciplinary team should assess, manage and review cognition and behaviour and should have prompt access to psychology/neuropsychology. It recommends exploring any cognitive or behavioural changes with the person and their family members and/or carers at diagnosis, and whenever there is concern about cognition and behaviour. If needed, the person should be referred for a formal assessment in line with the NICE guideline on dementia.<sup>67</sup>

Ideally, all patients should be referred to a neuropsychologist who undertakes an assessment such as the ECAS (see page 22-24). Where this is not possible, a member of the team should perform the assessment under the supervision of the neuropsychologist for interpretation.



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.<sup>53</sup>

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
- assessment of the persons abilities to undertake everyday activities
- standardised screening measure of cognition and behaviour
- informant-based behavioural interview questionnaires – these are separate interview with someone who knows well the person with MND which asks questions about possible changes in behaviour
- assessment according to current criteria for MNDbi (behavioural impairment), MNDci (cognitive impairment) and MND-FTD.<sup>8</sup>

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

## Screening tools

The Edinburgh Cognitive and Behavioural ALS Screen (ECAS) tool and the ALS Cognitive Behavioral Screen (ALS-CBS) are well-validated screening tools specifically designed for use in MND patients.<sup>54, 55, 62</sup>

These tools are brief and have been designed to minimise demands on speech or motor capacity. They can be completed by any health or social care professional, including non-neuropsychologists. They test cognitive and behavioural changes. Part of the assessments include questions for a carer about the behaviour of the person with MND.

### The ECAS tool

The ECAS tool has been designed specifically as a first step in assessing the presence of cognitive change in MND.<sup>54</sup> It is designed

to be completed in 20 minutes by a healthcare professional. It can be used by non-neuropsychology healthcare professionals, but training or supervision by a neuropsychologist is recommended.

It consists of a mini-assessment which tests functions that may be impaired in MND, including executive functions, language and fluency, as well as those not usually affected, such as memory and visuospatial skills. It includes a separate carer interview about behaviour, which is based on the diagnostic criteria for behavioural variant FTD. It shows good sensitivity and specificity to detect mild impairment compared with full neuropsychological evaluation<sup>56</sup> and is also sensitive to FTD and Alzheimer's Disease.<sup>66</sup>

More information can be found at <https://ecas.psy.ed.ac.uk/>

In order to undertake an ECAS it is strongly advised that you receive training and certification. Online training is available through the ECAS website, above. 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. As it is a comprehensive tool, specifically designed for MND, it may also form the core part of the neuropsychological assessment itself.

Given that cognitive and behavioural impairment may be hidden by physical disability, it is recommended to screen all patients.



- 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 be interpreted with the help of a neuropsychologist and discussed among the MDT.
- Performance on the ECAS by the person being screened may be affected by age and education.<sup>56</sup> People with poor schooling and/or reading or writing difficulties will do less well. This must be taken into consideration in the interpretation.<sup>57</sup>
- How the tool is used and how usage is supervised should be discussed as a multidisciplinary team.
- Beware of the label 'cognitive impairment' and what it means for the person with MND and their family. It should not affect equity of care.

### **ALS Cognitive Behavioral Screen (ALS-CBS)<sup>55</sup>**

ALS-CBS IS is a brief screening measure of cognition and behavior in people with MND. It is composed of two sections: cognitive and behavioral. The cognitive section can be administered by a physician or other clinical care staff. The behavioral section is completed by a carer, family member, or other informant. Find out more at [www.tandfonline.com/doi/abs/10.3109/17482961003727954](http://www.tandfonline.com/doi/abs/10.3109/17482961003727954)

**Note:** the research article will incur a cost, but the screening tool is available for free in the supplementary materials section.

### **Behavioural screening tools**

ECAS and ALS-CBS discussed above both have behavioural screening components. Two other behavioural screening tools - MiND-B and ALSFTD-Q can also be used to detect behavioural changes.

**MiND-B** is a simple tool for the identification and quantification of behavioural symptoms in ALS.<sup>58</sup> 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. Visit [www.neura.edu.au/research-centre/forefront/mind-tool](http://www.neura.edu.au/research-centre/forefront/mind-tool)



**ALSFTD-Q<sup>59</sup>** is an alternative behavioural screening tool for people with MND. It is a disease-specific questionnaire to detect behavioural changes in MND patients. As one of the symptoms of FTD is lack of insight, the questionnaire has to be filled in by a partner or a family member of the person with MND. Visit [www.alsftdq.nl](http://www.alsftdq.nl) for further information and to view the screening tool.

### **Neuropsychological assessment**

Many neuropsychologists undertake just an ECAS with their MND patients. Other methods commonly used include:

- interview
- detailed cognitive assessment – this involves a person completing a series of tasks which 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 effect it has on 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.

Challenges in terms of completing the assessment include:

- motor and speech impairments, which often mask cognitive difficulties and render it hard to carry out an assessment (note that the ECAS can be undertaken either speaking or in writing)
- 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
- diagnosis of cognitive impairment depends on evidence of executive dysfunction (including social cognition) and also possibly language dysfunction, so it is important to look at possible changes in all these domains<sup>8</sup> (eg language and behaviour)
- cognitive function can change over time, so reassessment should be considered. Parallel versions of the ECAS are available to avoid practice effects.<sup>4</sup>



# 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. They are also strongly associated with carer burden.<sup>60, 61</sup>

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
- before a decision is made on the use of non-invasive ventilation, a thorough assessment should be completed, including consideration of the person's capacity to make decisions and to give consent and whether the person is likely to accept treatment
- professionals may choose to give information that is simpler and more 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 diseases 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 to share

Information sheet 14A – *Advance Decision to Refuse Treatment (ADRT) and advance care planning*

*End of life guide*

### Information for you

*A professionals' guide to end of life care in MND booklet*

Download from [www.mndassociation.org/publications](http://www.mndassociation.org/publications) or contact MND Connect to order hard copies. Call 0808 802 6262 or email [mndconnect@mndassociation.org](mailto:mndconnect@mndassociation.org)

## Supporting carers and family members

Supporting carers is vital. Their individual needs may be complex, especially if cognitive and/or behavioural changes are severe in the person with MND.

Cognitive and behavioural changes are symptoms for which many carers feel unprepared, especially as they may not have been told that they can occur as part of MND. Clear explanations and instructions can help. The needs of carers and family members should be assessed, and support strategies advised.

In some cases where the need for respite is urgent, the situation 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.

# 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 paying attention, organising and planning, making decisions, spelling and/or behaviour problems such as apathy, loss of manners or loss of regard for others, 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. It may also interfere with relationships with family, carers and clinicians.
- 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 will have very mild problems, 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 (see pages 16-17).

## **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 FTD may lack mental capacity to make decisions. Capacity issues should be assessed where FTD is evident. Assessment is decision specific. 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 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, try to refocus attention or show them what to do.

**Help to problem-solve by:**

- refocusing their attention to 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, such as spelling 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 there are difficulties 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, or 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, or 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 or seeing things from another person's perspective
- support the carer/family in understanding the reasons behind what appears to be an inappropriate or uncaring response
- advise the carer/family that they should express their feelings verbally and as explicitly/simple as possible.

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 self-centredness is evident (eg loss of concern for carer/family):**

- support the 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 the 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 when someone is happy, sad, angry or frightened
- 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
- 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.

# Summary

All health and social care professionals should grasp the opportunity to empower and educate people with MND on cognitive and behavioural 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 it empowers them to help the person they care for.

Be aware of how you discuss and describe cognitive change and ensure that someone is not 'labelled'. 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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# How we can support you

## **MND Connect**

Our helpline offers help, information and support to people living with MND, carers, family and health and social care professionals.

**Email:** [mndconnect@mndassociation.org](mailto:mndconnect@mndassociation.org)

**Phone:** 0808 802 6262

## **Benefits Advice Service**

Qualified advisers can help identify benefits the person with MND and carers may be entitled to, and advise on how to claim. This service is confidential, impartial and free.

**[www.mndassociation.org/benefits-advice](http://www.mndassociation.org/benefits-advice)**

## **Information resources**

We produce high quality information resources for people living with MND, carers, family members and health and social care professionals.

**[www.mndassociation.org/publications](http://www.mndassociation.org/publications)**

## **MND Association website**

We have a wide range of information to support health and social care professionals working with people affected by MND.

**[www.mndassociation.org/professionals](http://www.mndassociation.org/professionals)**

## **Education**

Our education programme is designed to improve standards of care and quality of life for people living with and affected by MND.

**[www.mndassociation.org/education](http://www.mndassociation.org/education)**

## **MND Professionals' Community of Practice**

A peer led group of health and social care professionals encouraging and supporting the development of good care for people living with, or affected by, MND.

**[www.mndassociation.org/cop](http://www.mndassociation.org/cop)**

## **Support grants and equipment loan**

Where statutory provision is not available, we may be able to offer a support grant or loan equipment.

**[www.mndassociation.org/getting-support](http://www.mndassociation.org/getting-support)**

## Research into MND

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

[www.mndassociation.org/research](http://www.mndassociation.org/research)

## MND Register

The MND Register aims to collect information about every person with MND to help researchers focus their knowledge and expertise in the right areas.

[www.mndregister.ac.uk](http://www.mndregister.ac.uk)

## Regional staff

We have a network of regional staff with specialist knowledge of MND. They work closely with local statutory services and community care providers. Contact MND Connect for further information.

Email: [mndconnect@mndassociation.org](mailto:mndconnect@mndassociation.org)

Phone: 0808 802 6262

## MND care centres and networks

We fund and develop care centres and networks across England, Wales, and Northern Ireland, which offer specialist MND care.

[www.mndassociation.org/care-centres](http://www.mndassociation.org/care-centres)

## Branches and groups

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

[www.mndassociation.org/branchesandgroups](http://www.mndassociation.org/branchesandgroups)

## Association visitors (AVs)

AVs are trained volunteers who provide one-to-one local support to people affected by MND.

[www.mndassociation.org/associationvisitors](http://www.mndassociation.org/associationvisitors)

## We value your feedback

We would greatly appreciate your feedback on this guide. Please visit [www.smartsurvey.co.uk/s/mndprofessionals](http://www.smartsurvey.co.uk/s/mndprofessionals) or email your comments to [infofeedback@mndassociation.org](mailto:infofeedback@mndassociation.org)

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