



# Best Practice Guidelines for Health Care Professionals

Cognitive and behavioural changes in Motor Neuron Disease  
Prevalence, detection, and implications for diagnosis and management





## Executive Summary

Motor Neuron Disease (MND), (also known as Amyotrophic Lateral Sclerosis (ALS) is progressive motor system degeneration that presents in midlife and has a life expectancy of 3-5 years. Until recently, cognition was considered to be unaffected.

However, we now know that up to 40% of patients with MND also have mild to moderate cognitive and behavioural deficits and almost 15% of patients present with Fronto-temporal dementia.

The Edinburgh Cognitive and Behavioural ALS Screen (ECAS) is a short screening test for MND that can be administered by healthcare professionals. It assesses a wide range of cognitive domains and includes a short caregiver interview. A normal ECAS means that the patient is cognitively and behaviourally intact. However, those who score abnormally on the ECAS are not by definition impaired - they should be referred to Neuropsychology for comprehensive neuropsychological testing.

The diagnosis of cognitive and behavioural deficits in MND has important implications for individual patients and their families. These deficits may affect the person's capacity to make financial and health care related decisions.

Cognitive and behavioural deficits are also associated with reduced adherence to medical treatments and compliance with multidisciplinary care interventions. The ability to competently engage in end-of-life decisions can also be affected.

Cognitive and behavioural changes are a significant contributor to caregiver burden. Behavioural impairment in particular can be stressful for caregivers, as it can change the dynamics of relationships.

An understanding of components of caregiver burden leads to better support of the caregiver thus minimising the impact of the caregiver's mental health difficulties on the person with MND.

Healthcare professionals should be aware of the presence of cognitive and behavioural impairment in MND.

This booklet provides guidelines that aim to enhance clinical care in the context of complex physical, cognitive, behavioural and social changes that are associated with MND.

## Do All Patients With MND Develop Cognitive And Behavioural Changes?

Cognitive and behavioural deficits are now recognised as an important factor in MND. Within the last decade, evidence from neuropsychological, neuroimaging, neuropathological and genetic studies have established that cerebral involvement, which extends beyond the motor cortex, is implicated in MND.

Population-based studies of cognitive impairment in MND suggest that approximately 40% of patients present with mild to moderate cognitive and behavioural deficits. Although cognitive decline and behavioural change can co-exist in approximately 25% of MND patients, deficits can also occur in isolation. Cognitive and behavioural symptoms may precede, co-occur or follow the onset of the motor symptoms.

Additionally, up to 15% of patients with a diagnosis of MND also meet criteria for the diagnosis of Fronto-temporal Dementia (FTD).

FTD is a type of dementia that involves degeneration of frontal and temporal areas of the brain, and is characterised by changes in personality, behaviour and language. The combination of MND (or ALS) and FTD symptoms suggests the diagnosis of a widely accepted diagnostic entity known as ALS-FTD. That being said, not every patient with MND and cognitive and/or behavioural decline meets the diagnostic criteria for ALS-FTD.



## Which cognitive and behavioural changes do we observe most in MND patients?

The most common cognitive and behavioural changes reported in MND suggest the involvement of frontal and temporal areas of the brain, and are similar to a subtle form of FTD.

MND and FTD have been linked not only at a clinical and anatomical level, but also at a genetic and pathological level, with the discovery of common molecular dysregulations, including the RNA-binding protein FUS/TLS, TDP-43 or the hexanucleotide repeat expansion in the C9orf72 gene.

The frequency, type and progression of cognitive and behavioural changes in MND are very individual, and the severity and trajectory can range from very mild and subtle changes, to a more rapidly progressive clinical dementia.

### Cognition

The most common cognitive domain affected in MND is **executive function**. The term executive function refers to a set of higher-level cognitive skills such as attention/concentration, planning and organisation, mental flexibility, reasoning, problem-solving, self-monitoring and self-regulation. Patients with executive dysfunction may exhibit inattention or distractibility, difficulty planning and organising, mental rigidity or inflexibility characterised by decreased agreeableness with lowered frustration tolerance, and difficulty with problem solving. Judgement and decision-making may also be affected if executive dysfunction is present.

**Language** can also be impaired in MND, and often presents in the form of word-finding difficulties. Some patients may also have difficulties comprehending complex commands i.e., sequential commands, or with spelling. In more severe cases, individuals can lose the meaning of the words, not being able to recognise them or using them incorrectly in a sentence.

When MND patients present with **memory** deficits, these are in the form of reduced learning or poor encoding of new information. These patients may also have difficulty retrieving the information spontaneously. However, these deficits are often secondary to executive dysfunction and cues may facilitate the recall of the information learned, as storage is usually unaffected.

## Behaviour

Behavioural symptoms in MND are heterogeneous in nature. Some new behaviour tends to cluster together. The most prominent behavioural symptom reported in MND is apathy, which is defined as loss of motivation, initiative or interest. Individuals with **apathy** show passivity and lack of spontaneity. They may need to be prompted to initiate or continue routine activities or previously rewarding activities or hobbies. They may also manifest reduced interest in starting or sustaining conversation, and show less concern about self-care.

When evaluating the presence of apathy in MND, it is important to consider that motor disability and dysarthria may affect the patient's ability to initiate conversation or to engage with daily activities or previous hobbies that require motor involvement. Depression or fatigue due to respiratory dysfunction can also interfere with the patient's ability to engage in such activities.

**Disinhibition** or involuntary lack of verbal and physical self-restraint can also be observed in MND. This behaviour can manifest as loss of manners, socially inappropriate behaviour or difficulties with impulse control. Patients may display a general lack of social decorum, showing inappropriate behaviours such as

interrupting others in conversation, failing to wait in line, eating with mouth open, etc. Individuals can also exhibit inappropriate behaviour towards strangers such as improper approaching, touching or kissing.

Other behaviours that violate social graces such as inappropriate cursing, loudness, or rude or sexually explicit comments that may be offensive to others can be observed. More severe behaviours may include poor hygiene, impolite physical behaviours such as flatulence, belching or spitting, or aggressive physical or verbal behaviour. Disinhibited patients may be more irritable and prone to anger outbursts. Impulsivity is also frequently observed, which is associated with poor risk assessment.

Other more severe impulsive behaviours may include reckless driving, gambling, buying or selling objects without regard of consequences, and sharing private information such as credit card numbers.

**Loss of sympathy or empathy** can also be reported in MND, and it is characterised by lack of understanding or indifference to the needs and feelings of others. Individuals may also exhibit reduced interrelatedness or personal warmth, and family and friends may notice that the patient is emotionally detached and distant. A decline in social engagement may also be observed due to diminished social interest, which can lead to social withdrawal.

**Perseverative, stereotyped or compulsive behaviours** include simple repetitive movements (tapping, humming, rocking, rubbing, scratching, throat clearing, pursing of lips or lip smacking) or more complex ritualistic behaviours such as counting, ordering objects, cleaning rituals, collecting, hoarding or walking fixed routes. Patients may also consistently repeat single words, sentences or stories with no communicative value, which is known as stereotypy of speech.

Some patients experience **altered food preferences**. Although dietary changes are common due to swallowing difficulties, in this case the change in food choices manifest as stereotyped or idiosyncratic food preferences.

Some patients may exhibit food cravings, particularly of carbohydrates or sweets. Increased or new alcohol consumption or smoking can also be observed. In more severe cases, individuals may present with binge eating, which is persisting eating despite satiety, and in extreme cases, hyperorality can present as oral exploration, chewing or consumption of inedible items.

A **reduced awareness of their deficits and symptoms** can occur in more severe cases. This can range from a general lack of insight into the disease or a more specific **anosognosia**. Anosognosia is an inability to recognise the existence of the symptoms or disability associated with the disease. Anosognosia is not a deliberate or intentional denial on behalf of the patient. .

**Emotional lability** or **pseudobulbar affect** is very common in MND and it refers to episodes of spontaneous involuntary crying and/or laughing that can be inappropriate for the social situation. Although these episodes are not associated with an actual experience of the underlying emotion, they can be very distressing for the patient and their families. Emotional lability can occur independently of the presence of cognitive and other behavioural symptoms.

Finally, **psychotic symptoms** such as delusions or hallucinations have been observed in MND-FTD patients and are associated with the C9orf72 mutation.



## How to detect cognitive and behavioural changes in MND

A comprehensive neuropsychological assessment is required to definitively diagnose cognitive and behavioural change in MND, although it is recommended that patients are first screened using a disease specific assessment.

The Edinburgh Cognitive and Behavioural MND Screen (ECAS) is a short screening test for MND suitable for use in a clinic setting. This can be administered by trained healthcare professionals.

It largely accommodates for motor disability as it can be administered either in a written or spoken form.

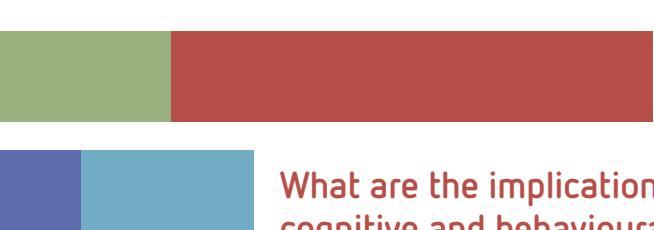
The ECAS takes approximately 15 minutes to complete and assesses a wide range of cognitive domains, including those more frequently affected in MND (MND Specific; executive function, lexical fluency; social cognition) and those not as frequently reported (MND Non-Specific; memory and visuospatial processing). It also includes a short caregiver interview to check for the presence of behavioural changes.

The ECAS is a sensitive measure for detection of cognitive deficits in the Irish MND population, although, there is a risk of generating false positives (i.e finding symptoms when none exist).

The ECAS does not take into account premorbid intellectual level, specific learning disability i.e., dyslexia, and behavioural status, and all patients who exhibit deficits on ECAS should be referred to a clinical neuropsychologist for a comprehensive neuropsychological assessment.

The ECAS can be downloaded from the Edinburgh Research Archive ([www.era.lib.ed.ac.uk](http://www.era.lib.ed.ac.uk)) and age and education matched Irish population specific cut-off scores are available upon request from the Irish MND/MND Research Group ([www.mnd.ie](http://www.mnd.ie))

Comprehensive neuropsychological assessments should be undertaken by appropriately trained psychologists. They should adhere to the current standard of care and include all appropriate cognitive domains paying particular attention to executive dysfunction, social cognition and language, as well as standardised carer-based interviews to assess behaviour.



## What are the implications of the diagnosis of cognitive and behavioural impairment in MND?

The diagnosis of cognitive and behavioural deficits in MND has important implications for individual patients and their families. Cognitive changes in MND have been associated with reduced survival. These deficits may also affect the patient's capacity to make financial and health care related decisions such as acceptance of gastrostomy insertion or need for non-invasive ventilation, and decisions about end of life. Safety awareness can also be affected. Cognitive and behavioural deficits are sometimes associated with reduced adherence to medical treatments and compliance with multidisciplinary care interventions.

Behavioural changes can affect interpersonal relationships, and contribute to caregiver burden. Caregivers should be provided with appropriate information regarding the types of cognitive and behavioural changes that can occur in association with MND

### Caregiver Burden

Caregiver burden is complex and multidimensional. However, changes in patient behaviour comprise a major factor in the level of caregiver burden and have a detrimental impact on their day-to-day life.

Deconstructing caregiver burden and recognition of the impact of cognitive and behavioural change on the relationship between the patient and caregiver can help healthcare practitioners to focus interventions.



## Managing cognitive and behavioural changes in MND

It is important to recognise that one of the main aspects of behavioural management in MND involves working with the carers and family on a multi-systemic level. This is based on helping to understand, accept and cope with new and distressing symptoms and behaviours. Education about the biological nature of these changes is crucial.

Attributing behavioural changes to brain pathology rather than a voluntary conduct of their loved ones helps carers in changing their emotional responses to these behaviours. This can help to change negative reactions to more adequate responses. For example, understanding that lack of insight, apathy or lack of empathy resulting from changes in brain pathology caused by MND, helps caregivers to shift expectations, and offers a more acceptable explanation as to why individual patients may not appreciate caregiver's tasks.

## Recommendations for management and care for patients with cognitive and behavioural deficits

### Care Planning

It is very important to explain clearly that cognitive and behavioural changes are part of the condition and to set realistic expectations for both patients and family members

It may not be possible for those with cognitive and behavioural impairment to comply with interventions such as non-invasive ventilation. Additionally, those with severe executive impairment or dementia may not be suitable for gastrostomy insertion. Some patients with ALS-FTD may exhibit swallowing difficulties due to changes in executive function rather than true dysphagia. These patients may not benefit from gastrostomy insertion.

Some patients may not be able to manage some alternative communication devices such as tablets, which are suitable for MND patients without cognitive and behavioural deficits.

Care plans for MND patients with cognitive and behavioural deficits may need to be simplified to be in line with the patient's current abilities. Treatment options and preferences may need to be discussed with the patient as soon as possible in the course of the disease. As some patients may lack proper ability to judge their care needs or may lack insight about their deficits, family members should be included in these discussions. Power of attorney should be discussed early in the course of the illness.

## **Advice for Families**

- Encourage the family to supervise walking and transfers, especially in the case of impulsive patients, to reduce the risk of falls.
- Advise that self-care and hygiene may need to be supervised.
- Encourage supervision during mealtimes, especially in the case of patients with swallowing difficulties. If impulsive eating is observed, encourage the patient to count the number of times the food is chewed before swallowing it. If food cravings or hyperorality are present, portions need to be limited and cupboards or refrigerators may need to be locked.
- When dealing with impulsive patients, items that may be a risk of harm for the patient need to be removed.
- Caregivers should be encouraged to provide a clear structure to the patient's day and make it as predictable as possible.
- Caregivers should be encouraged to give the patient enough time to make decisions and offer limited choices and closed ended questions. The "little and often" rule applies. Patients are more likely to understand and manage small amounts of information . Distractions should be minimised to assist with concentration.
- Caregivers should simplify communication to enhance patient's comprehension. They should be advised to speak clearly using a simple and straightforward language, to break sentences into short phrases containing not too much information, and slow down when speaking.
- Caregivers should encourage the patient to use calendars, memory aids or phone alarms
- In patients with increased irritability, caregivers should be advised to look for triggers that may prompt bursts of anger (tiredness, hunger, etc.) and try to prevent them. In cases of anger outburst, remain calm and avoid arguments, caregivers should be advised to acknowledge the patient's irritability.
- Caregivers should avoid surprises that may create confusion or agitation and keep the environment calm and controlled. If visitors are expected, the caregiver should make sure that the patient is aware of this. If large gatherings provoke agitation or irritability in the patient, avoid them.



## Caring for the Health Care Professional

Behavioural change in patients can be challenging for healthcare professionals (HCP). There are reports of heightened distress among professionals who care for those with MND and bvFTD. Because patients are disinhibited, they can be divisive, and can set one colleague against another.

This can often manifest from patients engaging in a 'splitting' process, whereby they engage with some professionals around certain tasks (e.g. will only let a particular staff member assist at meal-times) which can create stress and ill feeling within teams.

Patients with behavioural impairment are often unaware of their illness, and may be unable to appreciate the impact of the illness on others, or to appreciate the needs of others.

Some patients can appear manipulative, sexist, racist and abusive.

It can be extremely difficult for professionals to engage with patients and to develop appropriate care plans when there is reduced awareness, lack of insight, and when there are difficulties with problem solving and organising. This can lead to heightened stress for the professional.

Healthcare professionals should be empowered in dealing with these challenging behaviours, and while recognising the autonomy of the patient, the integrity of the professional must also be protected.

## **Recommendations**

- The HCP need to be honest in dealing with the patient, recognising that the illness may cause behavioural changes.
- The HCP should recognise that sometimes the behaviour is difficult and upsetting for people trying to provide care, and that they cannot always provide what the patient requests.
- The HCP should be firm, upfront in what services they can provide but also needs to ensure that the patient is included in the process, if practical.
- The HCP should avail of all support systems, including debriefing sessions when managing challenging behaviours.
- For patients with MND and bvFTD, care should be shared across a number of different HCPs within the community, or in a hospital /nursing home setting.
- In the community, carers of those with ALS-FTD should be rotated. The rational for this plan of care should be explained to the patient and caregiver, outlining that the disease can sometimes cause behavioural changes and that this behaviour is challenging for all concerned providing care.



## The Perspective of the Caregiver

- Behaviour changes pose more problems for families than cognitive changes
- Caregivers and families are in need of care also
- Caregivers need additional support from clinical and community teams who understand behavioural changes in MND
- Families want advice on how to manage the behavioural problems, and to learn coping strategies, as well as more information about the condition, how to cope with the future, and the opportunity to ask questions
- Families believe that it would be useful in managing behavioural changes to introduce formal 'outside' carers early on in the disease course, which might allow the patient to get used to the idea, and allow earlier support for the caregiver
- Families would like to have time and space to meet HCPs separately from patients, as they would be more comfortable to talk freely about their experiences in a sensitive manner without the patient being present;
- Networks and focus group meetings would be useful to enable information exchange
- It would be helpful for caregivers to be able to acknowledge their emotions to themselves and others

## Recommendations for provision of support for caregivers of MND patients with cognitive and behavioural deficits:

- Patients' and caregivers' perspectives might differ from those of the health professionals involved in their care.
- Effective caregiving requires that caregivers receive practical and emotional support.
- Routine cognitive-behavioural screening can identify patients who require full neuropsychological examination, and identify caregivers in need of early targeted interventions to assist in stress management and to manage caregiver burden.
- Identification of caregiver needs allows for the better management, leading to a reduction of overall burden and greater caregiver empowerment.
- MND caregivers need to become aware that patients may change their behaviour over time, and should be supported in the modification of their interpersonal relationship.
- To address the complex needs of caregivers, a detailed appraisal of the cognitive, behavioural and motor disability of the patient should be accompanied by detailed analysis of the psychological, emotional and capacity for resilience of the caregiver.
- Consideration of the possible positive aspects of caregiving is also important as support and interventions for caregivers' should also enhance the positive aspects of their role, which could reinforce their well-being and reduce the impact of caregiving in stress and burden.

This project was funded by a HRB Knowledge Exchange and Dissemination Award.

We are grateful to the patients, caregivers and families who have assisted in the development of these guidelines by permitting detailed cognitive, behavioural and caregiver burden assessments.

We are also grateful to the families who attended our town hall meetings and focus groups, and who provided us with important insights into the day to day problems associated with cognitive and behavioural changes in MND.