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Cognitive and Behavioural Changes in Motor Neuron Disease

A guide for families

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What are Cognitive and Behavioural Impairments in MND?

MND mostly affects the motor nerves which travel from the brain to the spine (upper motor neuron) and from the spine to the muscles (lower motor neuron). The degeneration of these groups of nerves can lead to difficulty with movement, speech, swallowing and breathing, leading to a diagnosis of MND.

We now know that some people with MND also experience degeneration in other parts of the brain, typically in the regions known as the frontal and temporal lobes. The frequency, type and progression of these changes in MND are very individual, and can range from very mild and subtle changes, to a more rapidly progressive dementia.

Cognition

The most common changes that occur are attention/concentration, planning and organisation, mental flexibility, reasoning, problem-solving, self-monitoring and/or self-regulation.

Patients may not be able to concentrate, and may have difficulty planning and organising. Judgement and decision-making may also be affected. This is usually a result of a combination of difficulties, such as future planning, problem-solving, and reasoning. Some people experience many of these changes, while others may experience only a few.

Language can also be affected in MND, often in the form of word-finding difficulties. Some patients may also have difficulties understanding complex sentences, and can make mistakes with spelling. In more severe cases, the person can lose meaning of the words, or may not be able to recognise them, or may use them incorrectly in a sentence. Some MND patients can also have

problems with their memory. This means that the person can sometimes forget things or events, but can remember them when prompted or after a while.

If you have noticed any such cognitive/thinking changes in your family member with MND, you should talk to your Neurologist and they can arrange a formal assessment.

Behaviour

Changes in behaviour can occur in some people with MND. In general, these changes are more noticeable than cognitive changes, and can be distressing to family members.

One of the most distressing behavioural changes for families and carers is the fact that some people with MND can become indifferent to the needs and feelings of others. Family and friends may notice that the person is emotionally detached and distant.

The most common change associated with MND is apathy. This is when the person loses motivation, initiative or interest. People with apathy seem more passive and lacking in spontaneity. They may need to be prompted to start or continue previously enjoyable activities or hobbies. They may also show less concern about self-care, beyond the physical limitations imposed by MND.

Some people with MND also have difficulty controlling their impulses. They can seem to have lost their manners, and they can start behaving inappropriately in company. They can appear rude by interrupting others in conversation, failing to wait in line, eating with mouth open, etc. Other behaviours can include inappropriate touching, or inappropriate cursing, loudness, or rude or sexually explicit comments that can be offensive to others. More severe changes in behaviour can include poor hygiene and impolite physical behaviours such as flatulence, belching or spitting. Patients affected with disinhibited and inappropriate behaviours are also more likely to take risks, and to ignore warnings. This might include reckless driving, gambling, buying or selling objects without regard of consequences, and sharing private information such as credit card numbers.

Disinhibited patients may also be less agreeable, more irritable and prone to anger outbursts, and physical or verbal aggressive behaviour can be observed at times due to a lower frustration tolerance.

New obsessional or compulsive behaviours can develop. These can include simple repetitive movements (tapping, humming, rocking, rubbing, scratching, throat clearing, pursing of lips or lip smacking), or more complex behaviours such as counting, ordering objects, cleaning rituals, collecting, hoarding or walking fixed routes. Patients may also consistently repeat single words, sentences or stories.

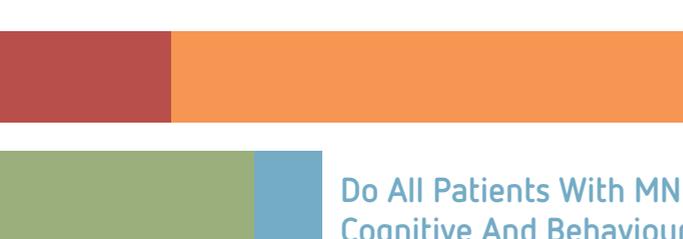
Some patients change the types of food that they prefer, and may have food cravings, particularly sweets or carbohydrates. In more severe cases, patients may begin to binge eat.

In more severe forms of behavioural change, the patient may not be aware of these changes, thus not being able to recognize them.

A small percentage of patients also experience new psychiatric symptoms, including delusions (i.e. feelings of persecution) or hallucinations (seeing/hearing things which are not present).

Inappropriate laughing or crying easily, known as emotional lability, is also very common in MND. This can be very distressing for the patient and their families but can be treated with medication.

When emotional lability is present, a person with MND may laugh or cry suddenly, without reason, intent, or the emotion associated with the act e.g., the person may cry but they are not sad/upset.



Do All Patients With MND Develop Cognitive And Behavioural Changes?

Not everybody with MND experiences these changes. Almost half of MND patients will not experience any cognitive/behavioural change throughout their illness. However, around 40% of people with MND experience some changes. Cognitive and/or Behavioural symptoms may precede, co-occur or follow physical symptoms of MND.

At the more severe end, approximately 15% of patients with a diagnosis of MND also develop dementia, resulting in severe changes in personality, behaviour and language.



How do we detect cognitive and behavioural changes in MND?

A short 20-minute paper-based screen has been devised to measure changes in thinking and behaviour in MND. This is administered by trained healthcare professionals in MND clinics. In cases where some degree of cognitive decline or behavioural change is suspected, a referral to a clinical neuropsychologist is made for more comprehensive assessment of behaviour and cognitive processes.



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

The diagnosis of cognitive and/or behavioural deficits in MND has important implications.

Cognitive changes in MND can affect a person's ability to make important decisions about their personal life, and about the types of treatments that might be on offer. Because the person may not appreciate the benefits of some treatments, they are sometimes less likely to use them.

Cognitive and behavioural changes can also lead to risk-taking behaviours such as going up and down stairs unaided, refusing to use walking aids or continuing to drive against medical advice. The person's awareness of risk of choking is also decreased.

Behavioural changes can affect interpersonal relationships, and contribute to stress within the family.



Other Implications

Legal Considerations

It is important to recognise if a person is at risk of not being able to make decisions about their possessions and assets. Unless somebody else has been identified who is legally entitled to make decisions on behalf of the person (this is called Power of Attorney), all of the assets and property are frozen and cannot be used by anyone else. This has serious implications.

Power of attorney can be put in place through a Solicitor. Any person who is in good mental health can nominate/appoint another individual to take actions on their behalf if they become incapacitated.

Power of attorney should be discussed at an early stage in MND, if the patient exhibits signs of cognitive or behavioural impairment.

Caregiver Burden

Those who care for people with MND with cognitive and/or behavioural impairment need support.

It is important for people who are providing care to recognise that changes in a person's thinking and behaviour can be a symptom of the illness. In such cases, it is crucial to keep in mind that these changes are part of the illness, and not a deliberate response of the person with MND.

Changes in the person's behaviour can be very stressful for the caregiver, and can lead to difficult emotions such as frustration, anger, guilt, anxiety or depression. These changes in personality often mean that the interpersonal relationship with the person with MND has changed, and it is important to acknowledge that.

Some people may benefit from mental health support services (counselling or psychotherapy) to explore their thoughts, feelings and reactions around this.



Managing cognitive and behavioural changes in MND

- Don't be afraid to raise your concern with the MND Team. If necessary, ask to see a member of the team without the patient being present, so that you can discuss your concerns openly.
- Consider speaking with a Solicitor about Power of Attorney.

Day to Day Management

- Use all of the available services provided by the community and IMNDA.
- Seek external help/care from the public health nurse – even if the patient objects.
- Provide a clear structure to the patient's day and make it as predictable as possible. Use calendars, memory aids or phone alarms.
- Simplify communication.
- Speak clearly using a simple and straightforward language. Break sentences into short phrases containing not too much information, and slow down when speaking.
- Give the patient enough time to make decisions. Offer limited choices and closed ended questions. Patients are more likely to understand and manage small amounts of information. Distractions should be minimised to assist with concentration.
- Supervise walking and transfers, especially in the case of impulsive patients, to reduce the risk of falls.
- Supervise self-care and hygiene.

- Mealtimes might need to be supervised, especially in the case of patients with swallowing difficulties.
- In patients with increased irritability, 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.
- Avoid surprises that may create confusion or agitation and keep the environment calm and controlled.
- If visitors are expected, make sure that the patient is aware of this. If large gatherings provoke agitation or irritability in the patient, avoid them.
- The Caregiver needs some "me" time i.e., time for themselves away from MND.

Caregivers need extra supports. The MND Clinic will work to develop these.

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