NHS Continuing Healthcare for motor neurone disease

Motor neurone disease (MND) is a progressive, terminal disease that attacks the motor neurones, or nerves, in the brain and spinal cord that control how the muscles work. This means that messages gradually stop reaching the muscles, leading to stiffness, weakness and wasting. MND can affect how people walk, talk, swallow, breathe and think. Not all symptoms happen to everyone, and it is unlikely they will all develop at the same time or in any specific order.[1]

Although the condition itself is unpredictable, the terminal prognosis is not. There is no cure for MND, and a third of people diagnosed with the disease die within a year and more than half within two years of diagnosis.[2] The focus of interventions is to manage symptoms to improve quality of life.

Due to the rapidly progressive nature of MND, it is highly likely that individuals with MND will have primary health-based needs at some point and reach the threshold for NHS Continuing Healthcare (CHC). This is a package of care arranged and funded solely by the NHS when an individual has been assessed as having a 'primary health need'.

Purpose of this guide

A report[32] by the Continuing Healthcare Alliance, of which the MND Association is a member, has found that the CHC system in England is failing people with MND and other conditions.

People are subjected to an exhausting, complicated and intimidating process which has led to inadequate care packages or being denied CHC altogether. It is estimated that only 18% of those screened for this funding are found to be eligible.[33] In some cases, the process has been so slow that by the time funding has been granted, the person with MND has already died, or died days later.

This booklet is designed to support health and social care professionals to better understand MND in the context of the CHC domains and enable professionals to support those who are eligible to access this vital support at what can be the most difficult time in their lives and when they need it the most.

This information sheet applies to England.

In Wales, local health boards are responsible for local health services and many of the rules for Continuing Healthcare that apply in England also apply in Wales, although there are some differences. Visit **www.wales.nhs.uk** for further information.

In Northern Ireland, there is no guidance on NHS Continuing Healthcare. However, health and social care services in Northern Ireland often follow NHS guidance, so a similar package of care may be offered. Contact the local health and social care trust for advice about what is available. Visit **www.hscni.net** to find local contact details.

If you live in Scotland, contact MND Scotland for advice. Visit **www.mndscotland.org.uk** for further information and support.

Information to share with people affected by MND

Information sheet 10D – *NHS Continuing Healthcare*

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

Beacon

Provides free consultations of up to 90 minutes with an independent NHS Continuing Healthcare adviser, funded by NHS England. This is only available in England. Their 'Navigational Toolkit' is designed to help people to navigate their assessment or appeal, and is available on their website.

Visit **www.beaconchc.co.uk** or call 0345 548 0300 for further information.

What is MND?

MND results in progressive muscle weakness and wasting, usually first affecting limbs, trunk or bulbar regions. MND can affect adults of any age, however incidence is highest in people aged 55-79.[3] It is insidious in its onset and development. The initial stages, speed and pattern of progression can vary from person to person.[4]

Symptoms of MND

Initially, MND can present as isolated and unexplained signs or symptoms, which may include:

- stumbling, falls or trips
- foot drop
- loss of dexterity
- weakened grip
- cramps
- fasciculations (involuntary contractions or twitching of groups of muscle fibers)
- change in voice quality, such as quiet or slurred speech
- awareness of swallowing changes, such as having to 'double swallow'
- muscle wasting.[3]

First symptoms may progress to:

- loss of function of limbs
- weakness and wasting of muscles of the trunk and neck
- progressive weakness of bulbar muscles, affecting speech, eating and drinking, and clearance of saliva[3] or mucus[5]
- respiratory symptoms such as breathlessness and hypercapnia (too much carbon dioxide in the blood)
- cognitive change or dementia.

The following factors, if present at diagnosis, are associated with shorter survival:

- speech and swallowing problems (bulbar presentation)
- weight loss
- poor respiratory function
- shorter time from first developing symptoms to time of diagnosis.[3]

Prognosis

A third of people with MND die within a year of diagnosis and more than half die within two years. For some people with MND, death can be very sudden, before an obvious end stage is reached. Others may experience a longer final stage, lasting many weeks. For most people with MND, death will be peaceful.[6] Cause of death is almost always respiratory failure as a result of muscle weakness and/or repeated chest infections, often due to aspiration.[1]

What is NHS Continuing Healthcare (CHC)?

NHS Continuing Healthcare (CHC) is a package of care for adults aged 18 or over which is arranged and funded solely by the NHS. In order to receive CHC funding, individuals have to be assessed by integrated care boards (ICBs) according to a legally prescribed decision-making process to determine whether they have a 'primary health need'.

It's principles and processes are laid out by the *National Framework for NHS Continuing Healthcare and NHS-Funded Nursing Care* (hereafter referred to as the National Framework) which was updated in 2022.

The decision-making process usually includes two phases:

1. NHS Continuing Healthcare Checklist

For most people, the first stage of the process is a screening tool known as the CHC Checklist. Completion of the checklist does not in itself indicate eligibility, only whether there is a need for a full assessment for CHC funding using a tool known as the Decision Support Tool (DST) - see next heading. An exception is if the person has 'a rapidly deteriorating condition that may be entering the terminal phase' – in which case the fast track pathway should be followed. See page 5.

Only the official CHC Checklist can be used. ICBs are not permitted to adapt it or use any other tools or methods to screen for CHC. They may move straight to a full assessment with the DST without screening using the checklist if they wish.

It should be completed by an appropriately trained health or social care practitioner, with completion of the checklist identified as part of their role. These professionals could include registered nurses, GPs, local authority staff such as social workers, care managers or social work assistants, depending on the local ICB's rules.

2. Decision Support Tool (DST)

If the CHC Checklist has been completed and indicates a full assessment, the ICB will arrange for a multidisciplinary team (MDT) to conduct a thorough assessment of the individual's health and social care needs, and their desired outcomes using an assessment tool known as the DST. It is used to investigate whether a person has a primary health need that would be eligible for CHC.

The National Framework defines a multidisciplinary team as either:

- two professionals who are from different healthcare professions, or
- one professional who is from a healthcare profession and one who is responsible for assessing individuals for community care services under section 47 of the National Health Service and Community Care Act 1990.

These are the minimum requirements. The National Framework states that the MDT conducting the assessment should comprise of professionals knowledgeable about the individual's care needs, and involved in their care, treatment or diagnosis.

People with MND often have a significant number of professionals involved in their care and support who can be called on to form part of the MDT. They can also offer supporting evidence and further information to support the completion of the DST.

The DST brings all of the evidence together in a single format to support consistent decision-making regarding eligibility and includes a recommendation as to whether the person is eligible for CHC. There is one single assessment process, and the same MDT members should be involved throughout, from the completion of domains to the recommendation discussion.

As the process of applying for CHC can be exhausting and confusing for people, we recommend that one key person is tasked with keeping the person with MND and their family updated and informed throughout the process.

Domains of care

The DST is designed to measure the level of a person's needs in 12 categories known as care domains:

- breathing
- nutrition
- continence
- skin
- mobility
- communication
- psychological and emotional needs
- cognition
- behaviour
- drug therapies and medication: symptom control
- altered states of consciousness
- any other significant needs.

Eligibility

Under each of the domains, needs are established under the following levels:

- no needs
- low
- moderate
- high
- severe
- priority.

Not all domains carry all six levels. For example, priority only applies to behaviour, breathing, drug therapies and medications, and altered states of consciousness. The DST provides descriptors for each level.

A clear decision of eligibility for NHS Continuing Healthcare would usually be expected in each of the following cases:

- a level of priority needs in any one of the four domains that carry this level **or**
- a total of two or more incidences of identified severe needs across all care domains.

In all cases, the overall need, and interactions between needs, should be taken into account, together with evidence from risk assessments, in deciding whether the person is eligible for CHC.

Eligibility recommendation

The MDT completing the DST make a recommendation on eligibility under four headings, known as key characteristics.

- **Nature:** the characteristics of the individual's needs, and the interventions required to meet those needs. It can be helpful to consider the impact of the need on the person's overall health and any particular skills, knowledge or training required to meet their needs.
- **Intensity:** the quantity, severity or continuity of needs. Consider how often interventions are required, and for how long. The number of care workers required at any one time and whether the care relates to needs over several domains should also be included if relevant.
- **Complexity:** the level of skills, knowledge or training needed to address an individual need or the range of needs and the interface between two or more needs. Consider how problematic it is to alleviate the needs and symptoms. How the needs interrelate and impact on each other is an important aspect to consider.
- **Unpredictability:** is about the degree to which the needs fluctuate and the challenges this creates. Consider whether the person is able to anticipate when their needs may arise, whether their condition is stable and how often the level of need changes.

The key characteristics above are extremely important in deciding someone's eligibility. They should be carefully considered where there are a number of domains with high and/or moderate needs, or severe level need combined with needs in a number of other domains.

The MDT must come together for a genuine and meaningful discussion before agreeing a recommendation regarding the person's eligibility. The ICB will make a final decision. They must verify the MDT's recommendation, and this may be via a decision-making panel.

This recommendation must be upheld by the ICB unless there are exceptional circumstances. The ICB may refer the case back to the MDT with instructions to carry out further work if the DST is not fully completed or there is inconsistency between the evidence provided and the recommendations. **The ICB cannot refuse to accept an MDT recommendation simply because they would have made a different recommendation when presented with the same evidence.**

Disputes and appeals[7]

If someone was screened out at the checklist stage:

The person may ask the ICB to reconsider its decision and agree to a full assessment of eligibility. The ICB should take all available information into account including from the individual, and their family and carers. The person should receive a clear, written response and be informed of their right to complain under the NHS complaints procedure if they are still dissatisfied.

If a full assessment has taken place using the DST:

This should be addressed through the local resolution procedures for the ICB. According to the National Framework, the person must write to the ICB within the time limit set by the local ICB - this is no longer set nationally. In some areas, this may be as short as two weeks. It is strongly recommended to discuss the timescale required with the individual ICB.

If the matter remains unresolved, the person may apply to the NHS England for an independent review of the decision, if they are unhappy with either:

- the procedure followed by the Board or a ICB in reaching its decision as to the person’s eligibility
- the decision regarding eligibility.

Fast Track Pathway Tool[7]

Fast track assessments should be used for people with rapidly deteriorating conditions who may be approaching end of life and need an urgent decision to be made regarding their eligibility.

This is not dependent on the person’s prognosis. There are still misconceptions among professionals that only those with a certain number of weeks prognosis are eligible. The National Framework states “*strict time limits that base eligibility on some specified expected length of life remaining should not be imposed: it is the responsibility of the appropriate clinician to make a decision based on the needs of the person*”.

The Fast Track Pathway Tool must be completed by an appropriate clinician, defined by the National Framework as a person who is:

- responsible for the diagnosis, treatment or care of the individual; **and**
- a registered nurse or a registered medical practitioner.

If you do not meet these criteria but believe the person with MND needs to be fast-tracked, you should contact the clinician responsible for their diagnosis, care or treatment, and ask them to consider completing the Fast Track Pathway Tool.

Fast-tracked recommendations for CHC should be accepted and actioned by the ICB immediately. If there are any concerns about use of the tool, the person’s care package should be delivered, and they should not experience any delays while this is addressed.

Once someone has been found eligible for CHC using the fast track tool, their funding should not be removed without a full review of their eligibility by an MDT using the DST.

MND in the context of the care domains

This section explores MND in the context of the care domains used in the NHS CHC Checklist and DST. It includes the DST descriptors for moderate, high, severe and priority levels of needs for your reference.

Please refer to the National Framework and DST for further information and descriptors for low levels of need.

Breathing

The cause of death in MND is almost always due to respiratory failure as a consequence of respiratory muscle weakness and/or repeated chest infections.[1]

Weakness in the diaphragm and accessory muscles causes respiratory insufficiency in most patients with MND. Bulbar muscle weakness contributes to various complications including increased risk of aspiration, weakened cough and abnormal respiratory patterns.

People with MND may experience hypercapnia (abnormally high levels of carbon dioxide in the blood) as a result of respiratory problems. Symptoms of mild hypercapnia include fatigue, headache, nightmares, confusion and excessive sleepiness. In severe hypercapnia, the person may experience disorientation, panic, hyperventilation, convulsions, unconsciousness, and eventually death.[8]

Ventilation

A person with MND may use a ventilator. If they use a non-invasive ventilator, they will need a carer available to allow repositioning of the mask if they are unable to use their arms. If the person with MND is ventilated via a tracheostomy, they will need support to care for the tracheostomy tube, including cleaning, replacing the tube and suctioning of fluid from the trachea.

The DST states under the severe category "*a non-invasive device to both stimulate and maintain breathing*." The vast majority of people who use NIV do not actually need it to stimulate a breath so arguably this statement doesn't quite fit. However, the other levels, do not capture the level of need a person with MND using NIV would require, so severe is the most appropriate level.

Some people with MND are unable to tolerate NIV due to saliva issues or bulbar weakness. It is important to stress that NIV was prescribed to reflect the person's level of needs, even if they are not using it.

People who are unable to breathe independently with invasive mechanical ventilation should be classed as a priority level of need.

Cough management

People with MND may not be able to cough effectively and will require support such as manual assisted cough, breath stacking or use of a mechanical insufflation/exsufflation machine (MI:E) sometimes known by the brand name CoughAssist.

Decision support tool descriptors

Moderate level of need:

- Shortness of breath or a condition which may require the use of inhalers or a nebuliser and limit some daily living activities.
- Episodes of breathlessness that do not consistently respond to management and limit some daily living activities.
- Requires low level oxygen therapy (24%), room air ventilators via a facial or nasal mask or other therapeutic appliances to maintain airflow where the individual can still spontaneously breathe e.g. CPAP (Continuous Positive Airways Pressure) to manage obstructive apnoea during sleep.

High level of need:

- Is able to breathe independently through a tracheotomy that they can manage themselves, or with the support of carers or care workers.
- Breathlessness due to a condition which is not responding to treatment and limits all daily living activities.

Severe level of need:

- Difficulty in breathing, even through a tracheotomy, which requires suction to maintain airway.
- Demonstrates severe breathing difficulties at rest, in spite of maximum medical therapy.
- A condition that requires management by a non-invasive device to both stimulate and maintain breathing (bilevel positive airway pressure, or non-invasive ventilation).

Priority level of need:

- Unable to breathe independently, requires invasive mechanical ventilation.

Further information for you

Information sheet P5 - *Evaluation and management of respiratory symptoms in MND*

Information to share with people affected by MND

Information sheet 8A – *Support for breathing problems*
Information sheet 8B – *Ventilation for MND*

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

Nutrition

Swallowing problems affect at least two-thirds of people with MND during the course of their illness.[9] When someone presents with this symptom, deterioration is likely to follow.

The person may experience:

- coughing
- episodes of choking
- recurrent chest infections
- aspiration and aspiration pneumonia due to inhaling food, drink and saliva.

Other symptoms related to MND can also cause weight loss and malnutrition include:

- depression
- cognitive impairment
- hypermetabolism
- anxiety
- respiratory insufficiency

- constipation
- fatigue
- physical difficulties buying, preparing and eating food.

People with MND may require skilled assistance at mealtimes to support with feeding, positioning and managing the risk of choking. Monitoring weight in MND is important, and use of the Malnutrition Universal Screening Tool (MUST) is useful as supporting evidence. Speech and language therapist (SLT) reports or guidance are also usual evidence because they detail the safest consistencies and what to monitor when eating and drinking. The SLT report would help to demonstrate that it is not necessarily straightforward eating and drinking assistance that the person requires.

Gastrostomy

As swallowing difficulties increase, the person may have a special diet or use alternative feeding methods including gastrostomy.

Depending on the effects of MND, they may need support with administering feeds and flushing out the tubes to prevent blockages. Daily observation and care of tube is important, and a care plan should be in place in case of accidental dislodging of the tube.

If the person declines a gastrostomy, or declines to be fed through it, they will be at a greater risk of aspiration and choking as their swallow continues to decline. They will need support to manage hunger and palliation of symptoms in this situation.

Decision support tool descriptors

Moderate level of need:

- Needs feeding to ensure adequate intake of food and takes a long time (half an hour or more), including liquidised feed.
- Unable to take any food and drink by mouth, but all nutritional requirements are being adequately maintained by artificial means, for example via a non-problematic PEG.

High level of need:

- Dysphagia requiring skilled intervention to ensure adequate nutrition/hydration and minimise the risk of choking and aspiration to maintain airway.
- Subcutaneous fluids that are managed by the individual or specifically trained carers or care workers.
- Nutritional status “at risk” and may be associated with unintended, significant weight loss.
- Significant weight loss or gain due to identified eating disorder.
- Problems relating to a feeding device (for example PEG) that require skilled assessment and review.

Severe level of need:

- Unable to take food and drink by mouth. All nutritional requirements taken by artificial means requiring ongoing skilled professional intervention or monitoring over a 24-hour period to ensure nutrition/hydration, for example I.V. fluids/total parenteral nutrition.
- Unable to take food and drink by mouth, intervention inappropriate or impossible.

Further information for you

Information sheet P8 - *Dysphagia*
Visit our webpages at **www.mndassociation.org/professionals/nutrition-and-enteral-feeding**

Information to share with people affected by MND

Information sheet 7A – *Swallowing difficulties*
Information sheet 7B – *Tube feeding*
Eating and drinking with MND – our guide to help people with MND adapt how they eat and drink, if needed. It includes information, tips and easy-swallow recipes.

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

Continence

While MND rarely affects bowel and bladder function, immobility may affect the ability to get to the toilet on time, and loss of muscle activity can affect the strength required to defecate. Additionally, people with a particular subtype of MND known as primary lateral sclerosis (PLS) may experience painful bladder spasms and urinary urgency.[10]

Respiratory problems in MND may present as excessive urination at night.[11]

Changes in eating, dehydration, anxiety, medication and poor mobility can cause constipation and faecal impaction. People with MND may experience reduced peristalsis and weakness of pelvic floor/abdominal muscles may also contribute. Diarrhoea may occur as a result of persistent constipation.[12]

Decision support tool descriptors

Moderate level of need:

- Continence care is routine but requires monitoring to minimise risks, for example those associated with urinary catheters, double incontinence, chronic urinary tract infections and/or the management of constipation or other bowel problems.

High level of need:

- Continence care is problematic and requires timely and skilled intervention, beyond routine care (for example frequent bladder wash outs/irrigation, manual evacuations, frequent re-catheterisation).

Skin integrity

Someone with MND will be aware that they need to turn or move but may need help to adjust their position. Good skin and pressure care is vital to prevent irritation, pressure sores and their associated complications such as infection and blood poisoning.[13]

The skin can become vulnerable to breakage due to malnutrition, weight loss, immobility and difficulties maintaining hygiene. Special care is required when moving or handling a person with MND.

Decision support tool descriptors

Moderate level of need:

- Risk of skin breakdown which requires preventative intervention several times each day without which skin integrity would break down.
- Pressure damage or open wound(s), pressure ulcer(s) with 'partial thickness skin loss involving epidermis and/or dermis', which is responding to treatment.
- An identified skin condition that requires a minimum of daily treatment, or daily monitoring/reassessment to ensure that it is responding to treatment.

High level of need:

- Pressure damage or open wound(s), pressure ulcer(s) with 'partial thickness skin loss involving epidermis and/or dermis', which is not responding to treatment
- Pressure damage or open wound(s), pressure ulcer(s) with 'full thickness skin loss involving damage or necrosis to subcutaneous tissue, but not extending to underlying bone, tendon or joint capsule', which is/are responding to treatment.
- Specialist dressing regime in place; responding to treatment.

Severe level of need:

- Open wound(s), pressure ulcer(s) with 'full thickness skin loss involving damage'
- Open wound(s), pressure ulcer(s) with 'full thickness skin loss with extensive or necrosis to subcutaneous tissue, but not extending to underlying bone, tendon or joint capsule' which are not responding to treatment and require regular monitoring/reassessment.
- Destruction and tissue necrosis extending to 'underlying bone, tendon or joint capsule or above'
- Multiple wounds which are not responding to treatment.

Mobility

General muscle tightness and spasticity can affect mobility and coordination, causing pain and increasing the risk of falls. Trips and falls are a common issue in MND, and a major cause of hospital admissions.

These issues can affect the torso, neck and the limbs.[4] As the condition progresses, most people with MND will need to use a wheelchair. Muscle weakness and wasting in MND leads to mobility issues and can affect balance and posture.

If the person can no longer assist or cooperate with transfers, this could move them from Moderate to High. Eventually, disability can progress to the extent that some people with MND are only able to move their eyes. See subheading *Locked-in state* in the *Communication* section.

Changes to mobility in MND can be very rapid so regular reassessment and review of the person's needs and the support provided is essential.

People with MND may experience oedema (fluid retention) due to restricted activity and posture. This can lead to:

- pain
- difficulty walking
- stiff muscles
- stretched and itchy skin
- tissue scarring
- opportunistic infection
- skin ulcers
- inelasticity of blood vessels
- joints and muscles
- decreased blood circulation to the affected areas.

Decision support tool descriptors

Moderate level of need:

- Not able to consistently weight bear.
- Completely unable to weight bear but able to assist or cooperate with transfers and/or repositioning.
- In one position (bed or chair) for the majority of time but is able to cooperate and assist carers or care workers.
- At moderate risk of falls (as evidenced in a falls history or risk assessment)

High level of need:

- Completely unable to weight bear and unable to assist or cooperate with transfers and/or repositioning.
- Due to risk of physical harm or loss of muscle tone or pain on movement needs careful positioning and is unable to cooperate.
- At a high risk of falls (as evidenced in a falls history and risk assessment).
- Involuntary spasms or contractures placing the individual or others at risk.

Severe level of need:

- Completely immobile and/or clinical condition such that, in either case, on movement or transfer there is a high risk of serious physical harm and where the positioning is critical.

Further information for you

View our webpages at **www.mndassociation.org/professionals/muscle-weakness**

Information to share with people affected by MND

Information sheet 11C – *Equipment and wheelchairs*

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

Communication

More than 80% of people with MND are affected by slurred, quiet speech, with some experiencing a complete loss of speech.[16] This is caused by spasticity, largely resulting in weakness of the tongue, lips, facial muscles, pharynx and larynx.[17] Weakened respiratory muscles can also reduce the volume of the voice, and give it a breathy quality.

People may communicate in a range of ways, including using augmentative and alternative communication (AAC). This could include low tech equipment, such as eye-pointing frames, alphabet charts or high tech portable electronic communication aids using computers or tablet devices. It takes time and skill to ascertain the person’s wishes and promote autonomy. It is important to consider and minimise risk, for example how to summon help and avoiding isolation.

Locked-in state

A person with MND’s condition can deteriorate to the point where they cannot move or communicate verbally due to complete paralysis of nearly all voluntary muscles in the body except for eye movements. The person will still be aware. Communication, although challenging, may be possible through eye movements or blinking.

Decision support tool descriptors

Moderate level of need:

- Communication about needs is difficult to understand or interpret or the individual is sometimes unable to reliably communicate, even when assisted. Carers or care workers may be able to anticipate needs through non-verbal signs due to familiarity with the individual.

High level of need:

- Unable to reliably communicate their needs at any time and in any way, even when all practicable steps to assist them have been taken. The individual has to have most of their needs anticipated because of their inability to communicate them.

Further information for you

View our webpages at **www.mndassociation.org/professionals/dysarthria**

Information to share with people affected by MND

Information sheet 7C – *Speech and communication support*

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

Psychological and emotional needs

Living with a terminal diagnosis such as MND can cause considerable psychological and emotional distress. The disease is characterised by a series of losses with the accompanying issues of grief and bereavement. Feelings of anxiety, depression and isolation are common,[18, 19,20] especially in the first year after diagnosis.[21] Concerns may include:

- fear of choking to death' or 'fighting for breath'
- loss of independence and dignity
- inability to cope and loss of control
- isolation and social withdrawal
- hopelessness.

Decision support tool descriptors

Moderate level of need:

- Mood disturbance, hallucinations or anxiety symptoms, or periods of distress, which do not readily respond to prompts, distraction and/or reassurance and have an increasing impact on the individual's health and/or well-being.
- Due to their psychological or emotional state the individual has withdrawn from most attempts to engage them in care planning, support and/or daily activities.

High level of need:

- Mood disturbance, hallucinations or anxiety symptoms, or periods of distress, that have a severe impact on the individual's health and/or well-being.
- Due to their psychological or emotional state the individual has withdrawn from any attempts to engage them in care planning, support and/or daily activities.

Information to share with people affected by MND

Emotional and psychological support booklet

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

Cognition

MND can affect the frontal and temporal areas of the brain, affecting thinking, reasoning and behaviour. Up to half of all people with MND experience some degree of cognitive change, ranging from mild effects to noticeable impairment. This increases to 80% in the final stages of the disease course.[22]

In those with MND affected by cognitive change, the most prominent impairment is typically in executive functions. These include setting and achieving goals, planning, problem-solving, and responding to new situations.

Some people also show changes in language, social cognition and behaviour (see also next heading).

Around 15% of people with MND will develop a type of dementia called frontotemporal dementia (FTD), which involves severe cognitive change.

People affected by FTD may experience difficulties with learning new tasks, social interactions, keeping safe, making decisions and managing their affairs or finances.[23] People with FTD may develop behavioural issues, including aggression. See next heading *Behaviour*.

It is important to be aware that the person with MND may not have insight into the changes to their cognition or behaviour. Involvement of an MND specialist in the MDT is important, as dementia nurses may not have an accurate understanding of the impact of frontotemporal dementia on the daily care needs of someone with MND.

Decision support tool descriptors

Moderate level of need:

- Cognitive impairment (which may include some memory issues) that requires some supervision, prompting and/or assistance with basic care needs and daily living activities. Some awareness of needs and basic risks is evident. The individual is usually able to make choices appropriate to needs with assistance. However, the individual has limited ability even with supervision, prompting or assistance to make decisions about some aspects of their lives, which consequently puts them at some risk of harm, neglect or health deterioration.

High level of need:

- Cognitive impairment that could, for example, include frequent short-term memory issues and maybe disorientation to time and place. The individual has awareness of only a limited range of needs and basic risks. Although they may be able to make some choices appropriate to need on a limited range of issues, they are unable to consistently do so on most issues, even with supervision, prompting or assistance. The individual finds it difficult even with supervision, prompting or assistance to make decisions about key aspects of their lives, which consequently puts them at high risk of harm, neglect or health deterioration.

Severe level of need:

- Cognitive impairment that may, for example, include, marked short or long-term memory issues, or severe disorientation to time, place or person.
- The individual is unable to assess basic risks even with supervision, prompting or assistance, and is dependent on others to anticipate their basic needs and to protect them from harm, neglect or health deterioration.

Further information for you

Cognitive change, frontotemporal dementia and MND

Information to share with people affected by MND

Changes to thinking and behaviour with MND

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

Behaviour

Behavioural impairment is a recognised feature of MND. Research has shown that behavioural changes such as apathy, disinhibition and stereotypical behaviour are pervasive.[24]

It is important to be aware that this domain does not only refer to aggressive or violent behaviour. The National Framework explains that 'challenging behaviour' in this domain includes, but is not limited to:

- aggression, violence or passive non-aggressive behaviour
- severe disinhibition
- intractable noisiness or restlessness
- resistance to necessary care and treatment (but not including situations where an individual makes a capacitated choice not to accept a particular form of care or treatment offered.)
- severe fluctuations in mental state
- inappropriate interference with others
- identified high risk of suicide.

Behavioural problems in MND may include:

- behavioural disinhibition – socially inappropriate behaviour, disinhibited comments, loss of manners
- acting impulsively without thinking, inability to delay gratification (may include gambling, inappropriate internet spending, buying from cold callers etc)
- apathy and inertia – being withdrawn and distant, lacking interest, not initiating activities[25, 26]
- loss of sympathy and empathy for others – reduced response to other people's needs and feelings and social cues
- 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. Cramming food can be especially dangerous if the person has bulbar weakness due to the increased risk of choking.

Professionals must be aware of the risks to carers and family where behaviour changes include aggression. A combination of lack of empathy and self-seeking behaviour associated with FTD may lead to carers and family or the person themselves being in danger, especially if the person has retained mobility.

Note: People who appear not to be affected by cognitive change can nonetheless have profound behavioural abnormalities.

Decision support tool descriptors

Moderate level of need:

- 'Challenging' behaviour that follows a predictable pattern. The risk assessment indicates a pattern of behaviour that can be managed by skilled carers or care workers who are able to maintain a level of behaviour that does not pose a risk to self, others or property. The individual is nearly always compliant with care.

High level of need:

- 'Challenging' behaviour of type and/or frequency that poses a predictable risk to self, others or property. The risk assessment indicates that planned interventions are effective in minimising but not always eliminating risks. Compliance is variable but usually responsive to planned interventions.

Severe level of need:

- 'Challenging' behaviour of severity and/or frequency that poses a significant risk to self, others or property. The risk assessment identifies that the behaviour(s) require(s) a prompt and skilled response that might be outside the range of planned interventions.

Priority level of need:

- 'Challenging' behaviour of a severity and/or frequency and/or unpredictability that presents an immediate and serious risk to self, others or property. The risks are so serious that they require access to an immediate and skilled response at all times for safe care.

Further information for you

Cognitive change, frontotemporal dementia and MND

Information to share with people affected by MND

Changes to thinking and behaviour with MND

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

Drug therapies and medication: symptom control

A person with MND may require physical assistance to take medications, including administration via gastrostomy, syringe driver, patches, mouth, or nebuliser. A person with cognitive change or FTD may require additional support and prompts to take their medication as prescribed.

The potential complexity of medication given to someone with MND could create side effects, have variable effectiveness or create fluctuation of their condition or mental state, which should be reflected in the assessment.

A person with MND may have been prescribed anticipatory medication, which needs to be administered in response to sudden onset of distressing symptoms as opposed to set times.

Pain

This domain also considers pain, which can be an issue for many people with MND. Pain in MND may be caused by:

- muscle cramps
- spasticity
- mechanical stress on joints from muscle weakness
- skin sensitivity
- immobility
- oedema
- constipation.

People with MND may feel discomfort and pain, but be unable to move to get comfortable, so support may be required. Particular care is required when moving and handling someone with MND.

Decision support tool descriptors

Moderate level of need:

- Requires administration of medication (by a registered nurse, carer or care worker) due to non-compliance, type of medication or route of medication (for example PEG).
- Moderate pain which follows a predictable pattern; or other symptoms which are having a moderate effect on other domains or on the provision of care.

High level of need:

- Requires administration and monitoring of medication regime by a registered nurse, carer or care worker specifically trained for the task because there are risks associated with the potential fluctuation of the medical condition or mental state, or risks regarding the effectiveness of the medication or the potential nature or severity of side-effects. However, with such monitoring the condition is usually non-problematic to manage.
- Moderate pain or other symptoms which is/are having a significant effect on other domains or on the provision of care.

Severe level of need:

- Requires administration and monitoring of medication regime by a registered nurse, carer or care worker specifically trained for this task because there are risks associated with the potential fluctuation of the medical condition or mental state, or risks regarding the effectiveness of the medication or the potential nature or severity of side-effects. Even with such monitoring the condition is usually problematic to manage.
- Severe recurrent or constant pain which is not responding to treatment.
- Non-compliance with medication, placing them at risk of relapse.

Priority level of need:

- Has a drug regime that requires daily monitoring by a registered nurse to ensure effective symptom and pain management associated with a rapidly changing or deteriorating condition.
- Unremitting and overwhelming pain despite all efforts to control pain effectively.

Further information for you

Information sheet P11 - *Pain in MND*

Information to share with people affected by MND

Information sheet 6C - *Managing pain*

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

Altered states of consciousness

This domain relates to any condition which is significantly different from a person's normal waking state. This could include strokes, seizures and transient ischaemic attacks.

With MND, some people with respiratory failure, or at the end stage of life might be less responsive and difficult to rouse, and therefore at greater risk of unmet needs in terms of nutrition, hydration, hygiene etc. Those on strong opiates may also be less responsive.

Decision support tool descriptors

Moderate level of need:

- Occasional (monthly or less frequently) episodes of ASC that require the supervision of a carer or care worker to minimise the risk of harm.

High level of need:

- Frequent episodes of altered states of consciousness that require the supervision of a carer or care worker to minimise the risk of harm.
- Occasional altered states of consciousness that require skilled intervention to reduce the risk of harm.

Priority level of need:

- Coma.
- Altered states of consciousness that occur on most days, do not respond to preventative treatment, and result in a severe risk of harm.

Other significant care needs to be taken into consideration

This domain is designed to capture needs that do not relate to any of the other care domains. This could include other conditions the person may have related to MND, or due to another health condition. common issues in MND not covered by the other domains are explained below.

Fatigue

Many people with MND experience fatigue, contributing to a poor quality of life. Increased muscle weakness, wasting and problems with mobility will lead to muscles tiring more quickly. This, combined with other metabolic changes, can lead to fatigue. It can affect people in different ways but is usually experienced as an overwhelming tiredness that occurs gradually or suddenly, often not in proportion to the activities being undertaken. Fatigue tends to worsen throughout the day and is only partially relieved by rest.[30]

Fatigue in MND can be worsened by other symptoms of the disease such as low mood, sleep problems, inadequate nutrition, breathlessness and carbon dioxide retention (hypercapnia).

Further information for you

Occupational therapy for MND

Information to share with people affected by MND

Information sheet 11D - *Managing fatigue*

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

Emotional lability

25-50% of people with MND experience emotional lability (also known as pseudobulbar affect) which occurs as a consequence of upper motor neurone involvement in the corticobulbar tracts. This results in uncontrollable laughter or crying in response to something that is only moderately funny or sad.

There may also be inappropriate responses (eg laughing at a funeral or when in pain). This can be disturbing to both the person and those around them. For some, it limits where they go and what they do, leading to withdrawal and isolation.

Information to share with people affected by MND

Information sheet 9C - *Managing emotions*

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

Saliva management

Excessive saliva is a commonly reported symptom of MND. The amount of saliva produced is usually normal, and problems usually result from poor lip seal and an impaired ability to swallow.

People with MND may experience runny, thin saliva that drools out of the mouth, or thick, tenacious saliva that is difficult to clear. Problems with saliva may be managed using medications, nebulisers, radiotherapy, positioning and use of oral suction units. This symptom can be challenging to manage, and can impact on other aspects of MND, such as using NIV and staying hydrated and nourished.

Further information for you

Information sheet P3 - *Managing saliva problems in MND*

Information to share with people affected by MND

Information sheet 7A - *Swallowing difficulties*

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

Key points

- MND is a rapidly progressing and ultimately fatal disease.
- Many people with MND are having difficulty accessing the CHC support they are entitled to.
- MND symptoms affect all of the care domains within the CHC National Framework.
- People with MND do not have time to waste. Fast-tracking should be considered for those with rapidly progressing symptoms to ensure support is timely.
- The MND Association can offer additional information and support to people with MND and health and social care professionals. Email **mndconnect@mndassociation.org** or call 0808 802 6262.

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How we can support you and your team

MND Connect

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

Email: mndconnect@mndassociation.org
Phone: 0808 802 6262

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

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

Education

Our education programme is designed to improve standards of care and quality of life for people living with and affected by MND. Opportunities include online modules and face-to-face training.

www.mndassociation.org/education

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

Research into MND

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

www.mndassociation.org/research

MND register

The MND Register of England, Wales and Northern Ireland aims to collect information about every person living with MND to help plan care and discover more about the cause of the disease.

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
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 multidisciplinary care for people with MND.

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

Association visitors (AVs)

AVs are trained volunteers who provide one-to-one local support to people affected by MND. They can support people affected by MND in person, by telephone or by email or through support groups.

www.mndassociation.org/associationvisitors

We value your feedback

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

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

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