MND Association | For professionals

NHS

P7

NHS Continuing Healthcare for Motor Neurone Disease

Motor Neurone Disease (MND) results from the progressive loss of motor neurones in the brain and spinal cord. These are the nerve cells that control movement. It leads to muscle weakness and can cause difficulties with movement, breathing, swallowing and speaking.

Although the course of MND is unpredictable, it is a terminal condition. 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. The focus of interventions is to manage symptoms to improve quality of life.

Due to the progressive nature of motor neurone disease (MND), many individuals develop complex and increasing care needs over time. Some people with MND may reach the threshold for NHS Continuing Healthcare (CHC), following assessment.

CHC is a package of care arranged and funded solely by the NHS for individuals who have been assessed as having a primary health need. Eligibility is determined through assessment using the National Framework and is based on the nature, intensity, complexity and unpredictability of a person's needs. A diagnosis of MND alone does not determine eligibility.

Information to share with people with or affected by MND:

Information sheet 10D – NHS Continuing Healthcare

See page 26 for details of how to order publications.

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

1. Introduction

A report 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.[1]

People are subjected to an exhausting, complicated and intimidating process which has led to inadequate care packages or being denied CHC altogether. Around one in five people who undergo a full NHS Continuing Healthcare assessment are found eligible.[2] 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 **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 **hscni.net** to find local contact details.

In Scotland, NHS Continuing Healthcare has been replaced by Hospital Based Complex Clinical Care. Support for people with complex needs is provided through integrated health and social care services rather than a separate CHC process. Contact the local health and social care partnership for advice about available support. Visit MND Scotland at **mndscotland.org.uk** for further guidance.

2. What is MND?[3, 4]

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 55-75. It is insidious in its onset and development. The initial stages, speed and pattern of progression can vary from person to person.

Symptoms of MND

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

- stumbling, trips or falls
- foot drop
- loss of dexterity or weakened grip
- muscle cramps
- fasciculations (involuntary twitching of muscle fibres)
- change in voice quality, such as quiet or slurred speech
- awareness of swallowing changes, such as needing to ‘double swallow’
- muscle wasting.

As the disease progresses, symptoms may include:

- progressive weakness and loss of limb function
- weakness and wasting of the trunk and neck muscles
- bulbar weakness affecting speech, eating and drinking, and clearance of saliva or mucus
- respiratory symptoms such as breathlessness and hypercapnia (raised carbon dioxide levels in the blood)
- cognitive change or frontotemporal dementia.

The following factors, if present at diagnosis, are associated with shorter survival:[5, 6]

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

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.

Cause of death is most commonly respiratory failure as a result of muscle weakness and/or repeated chest infections, often due to aspiration.

3. What is NHS Continuing Healthcare (CHC)?[7]

NHS Continuing Healthcare (CHC) is a package of care for adults aged 18 or over, 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’.

The 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.

Phase 1: NHS Continuing Healthcare Checklist

For most people, the first stage of the process is a screening tool known as the CHC Checklist, which indicates whether a person should receive a full assessment using the Decision Support Tool (DST) (see next heading). An exception is where the person has ‘a rapidly deteriorating condition that may be entering the terminal phase’. In these cases the Fast Track pathway should be used instead (see page 7).

Only the official CHC Checklist can be used. Integrated Care Boards (ICBs) cannot modify it or use alternative screening tools, although they may proceed directly to a full DST assessment if appropriate.

The checklist should be completed by an appropriately trained health or social care practitioner, such as a registered nurse, GP, or local authority professional (for example a social worker or care manager), depending on local ICB arrangements. The ICB is responsible for ensuring appropriate access to the checklist locally. Any difficulties obtaining one should be raised with the relevant ICB CHC team.

Practitioners review each domain and select A, B or C, choosing the level that best reflects the person’s needs. If needs are equivalent to or greater than those described in column A, level A should be selected. A brief summary of the person’s needs and supporting evidence should be recorded.

A full DST assessment should be undertaken if the checklist identifies:

- two or more domains scored A
- five or more domains scored B, or one A and four Bs
- one A in a domain marked with an asterisk (breathing, behaviour, drugs and medication or altered states of consciousness), regardless of other scores.

In rare cases, a full assessment may still be appropriate even if these thresholds are not met. The reasons should be clearly documented and local protocols followed.

Phase 2: Decision Support Tool (DST)

If the CHC Checklist indicates that a full assessment is required, the Integrated Care Board (ICB) will arrange for a multidisciplinary team (MDT) to assess the individual's health and social care needs using the Decision Support Tool (DST). The DST is a tool used to determine whether a person has a primary health need and may therefore be eligible for CHC.

The National Framework defines an MDT as either:

- two professionals from different healthcare professions, or
- one healthcare professional and one professional responsible for assessing community care needs under the Care Act 2014.

These are the minimum requirements. The MDT should also include professionals involved in the person's care who understand their needs. For people with MND, this may include evidence from occupational therapists (for example manual handling assessments), speech and language therapists, and respiratory specialists.

The DST brings together all relevant evidence in a structured format to support consistent decision making and includes an MDT recommendation on CHC eligibility. The same MDT members should normally be involved throughout the assessment process.

As the CHC process can be complex and demanding, it is helpful for one professional to keep the person with MND and their family informed and updated throughout.

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 (behaviour, breathing, drug therapies and medications, and altered states of consciousness) **or**
- two or more incidences of severe needs identified across all care domains.

In all cases, the overall nature, intensity, complexity and unpredictability of the person's needs, and how these interact, should be considered alongside evidence from risk assessments when determining eligibility for CHC.

Eligibility is not dependent on having a priority level or two severe levels of need. These represent the upper threshold in the Decision Support Tool, but individuals with lower levels of need may still be eligible depending on their overall needs.

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 central to determining eligibility for CHC. They should always be considered alongside the domain scores, as they help to explain the nature, intensity, complexity and unpredictability of the person's needs and how these interact.

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

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 using the DST. 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 of the ICB. According to national guidance, the individual should write to the ICB within six months of the date of the decision letter if they wish to request a review of the eligibility decision. It is advisable to contact the relevant ICB CHC team for information about the local review process.

If the matter remains unresolved, the person may apply to NHS England for an independent review of the decision, if they are unhappy with either the procedure followed by the Board or an ICB in reaching its decision as to the person's eligibility or the decision regarding eligibility.

Fast Track Pathway Tool

Fast track assessments should be used for people with rapidly deteriorating conditions who may be approaching the end of life and need an urgent eligibility decision. When describing someone's MND as rapidly deteriorating, it is helpful to show week-by-week changes, detailing what the person could do one week but not the next.

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.

4. 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[8, 9, 11]

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

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 and aspiration pneumonia, 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.

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." Most 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 may be useful to state that NIV would have been prescribed, but the person was unable to use it to highlight the level of the person's needs.

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 Airway 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.

Information for professionals:

Information sheet P5 – Evaluation and management of respiratory symptoms in MND

Information to share with people with or affected by MND:

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

See page 26 for details of how to order publications.

Nutrition[8, 9, 12]

Swallowing problems affect at least two-thirds of people with MND during the course of their illness. 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 MND symptoms that 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 nutritional status in MND is important, and use of local screening tools such as MUST are useful to guide referral to dietitians, who will carry out detailed nutritional assessment and reviews, which could be used as supporting evidence. These would help demonstrate the main symptoms of MND contributing to nutritional decline.

Speech and language therapist (SLT) reports or guidance are also useful 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[12]

As swallowing difficulties increase, or nutritional status or weight decline, 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 the tube is important, and a care plan should be in place in case it is dislodged.

If the person declines a gastrostomy, or declines to be fed through it, they will be at 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.

Information for professionals:

Information sheet P8 – Managing dysphagia in MND

Information to share with people with or affected by MND:

Information sheet 7A – Swallowing difficulties
Information sheet 7B – Tube feeding
Booklet – Eating and drinking with MND

See page 26 for details of how to order publications.

Continence[8, 9, 13]

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. People with a particular subtype of MND called primary lateral sclerosis (PLS) may also experience bladder problems, most commonly urinary urgency, increased frequency and urinary retention.

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

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.

An inability to communicate or a locked-in state, whether due to physical disability or FTD, may prevent a person from expressing the need to urinate or pass faeces. Although this may not meet the clinical definition of incontinence, the person is effectively incontinent.

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[14]

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

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 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[8, 9]

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. 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.

Information for professionals:

Web page: **mndassociation.org/professionals/muscle-weakness**

Information to share with people with or affected by MND:

Information sheet 11C – Equipment and wheelchairs

See page 26 for details of how to order publications.

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 .

Communication[8, 10]

More than 80% of people with MND are affected by slurred, quiet speech, with some experiencing a complete loss of speech. This is caused by spasticity, largely resulting in weakness of the tongue, lips, facial muscles, pharynx and larynx. 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 may include low-tech tools such as eye-pointing frames or alphabet charts, or high-tech electronic communication aids using computers or tablet devices.

It takes time and skill to ascertain the person’s wishes and support autonomy. It is important to consider and minimise risks, for example how the person can summon help and avoid isolation. Impaired communication may also affect other areas of care if the person is unable to express needs or distress in a timely or effective way.

Locked-in state[10]

A person with MND can deteriorate to the point where they cannot move or communicate verbally due to paralysis of nearly all voluntary muscles in the body except for eye movements. The person remains 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.

Information for professionals:

Booklet – Communication, speech and language support

Information to share with people with or affected by MND:

Information sheet 7C – Speech and communication support

See page 26 for details of how to order publications.

Psychological and emotional needs[8, 9]

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, especially in the first year after diagnosis. 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 with or affected by MND:

Booklet – Emotional and psychological support

See page 26 for details of how to order publications.

Cognition[15, 16]

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.

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. 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.

Information for professionals:

Booklet – Cognitive change, frontotemporal dementia and MND

Information to share with people with or affected by MND:

Booklet – Changes to thinking and behaviour with MND

See page 26 for details of how to order publications.

Behaviour[9, 15]

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

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 issues in MND may include:

- behavioural disinhibition – socially inappropriate behaviour, disinhibited comments, loss of manners
- impulsivity – acting without thinking or delaying gratification (for example gambling or inappropriate spending)
- apathy and inertia – withdrawal, lack of interest, and reduced initiation of activities
- loss of sympathy or empathy – reduced response to others' needs, feelings or social cues
- perseverative, rigid or compulsive behaviour – repetitive movements or phrases, repeated responses, checking or hoarding
- hyperorality and dietary change – overeating or cramming, altered food preference (often sweet foods), excessive drinking or smoking. Cramming can be particularly dangerous with bulbar weakness due to increased choking risk.

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.

Information for professionals:

Booklet – Cognitive change, frontotemporal dementia and MND

Information to share with people with or affected by MND:

Booklet – Changes to thinking and behaviour with MND

See page 26 for details of how to order publications.

Drug therapies and medication: symptom control[8]

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[17]

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 (e.g. 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.

Information for professionals:

Information sheet P11 – Pain in MND

Information to share with people with or affected by MND:

Information sheet 6C – Managing pain

See page 26 for details of how to order publications.

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, vasovagal syncope (fainting) and transient ischaemic attacks.

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[8, 18]

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.

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).

Emotional lability[15]

25 to 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 (for example, 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.

Saliva management[8]

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.

Reduced responsiveness[8]

With MND, some people with respiratory failure or in the later stages of the disease may become less responsive and difficult to rouse, placing them at greater risk of unmet needs, such as nutrition, hydration and hygiene. Those on strong opiates may also be less responsive.

References

1. Continuing Healthcare Alliance. ‘Continuing to care? Is NHS continuing healthcare supporting the people who need it in England?’ London: Continuing Healthcare Alliance; 2016.
2. Hutchings R, et al. ‘All or nothing? Access and variation in NHS continuing healthcare.’ London: Nuffield Trust; 2025.
3. Hobson EV, et al. ‘Clinical aspects of motor neurone disease.’ Medicine (Abingdon). 2016;44(9):552–556.
4. Dharmadasa T, et al. ‘Motor neurone disease: progress and challenges.’ Med J Aust. 2017;206(8):357–362.
5. Motataianu A, et al. ‘Clinical and paraclinical predictors of survival in amyotrophic lateral sclerosis.’ Diseases. 2025.
6. Nakayama Y, et al. ‘Body weight variation predicts disease progression in amyotrophic lateral sclerosis.’ Sci Rep. 2019.
7. Department of Health and Social Care. ‘National framework for NHS continuing healthcare and NHS-funded nursing care.’ Revised ed. London: DHSC; 2022.
8. National Institute for Health and Care Excellence. ‘Motor neurone disease: assessment and management (NG42).’ London: NICE; 2016.
9. Masrori P, et al. ‘Amyotrophic lateral sclerosis: a clinical review.’ Eur J Neurol. 2020;27:1918–1929.
10. Ball LJ, et al. ‘Augmentative and alternative communication for people with ALS.’ J Commun Disord. 2018.
11. Lechtzin N, et al. ‘Respiratory care in amyotrophic lateral sclerosis.’ Chest. 2018;154:703–708.
12. Stavroulakis T, et al. ‘Management of nutrition in ALS.’ J Neurol. 2016;263:1789–1796.
13. Statland JM, et al. ‘Primary lateral sclerosis.’ Neurol Clin. 2015;33(4):749–760.
14. National Institute for Health and Care Excellence. ‘Pressure ulcers: prevention and management (CG179).’ London: NICE; 2014.
15. Strong MJ, et al. ‘Amyotrophic lateral sclerosis–frontotemporal spectrum disorder (ALS-FTSD): revised diagnostic criteria.’ Amyotroph Lateral Scler Frontotemporal Degener. 2017.
16. Mioshi E, et al. ‘Clinical staging and disease progression in amyotrophic lateral sclerosis.’ Neurology. 2016;87:1012–1018.
17. Chiò A, et al. ‘Pain in amyotrophic lateral sclerosis.’ Lancet Neurol. 2017;16:144–157.
18. Lou JS et al. ‘Fatigue and depression are associated with poor quality of life in ALS.’ Neurology. 2003;60(1):122–123.

How we can support you and your team

Our MND Connect helpline offers practical and emotional support, information and signposting to people with MND, carers, family and professionals. They can also provide further information about our services mentioned below.
Email: **mndconnect@mndassociation.org**
Phone: **0808 802 6262**

MND Association website

Our website offers supporting information on MND, our work, services, and how to get involved.
mndassociation.org/professionals

Stay updated on events, publications and opportunities for professionals.
mndassociation.org/educationupdate
X: **mndeducation**
Bluesky: **mndeducation.bsky.social**

Information resources

We produce high quality information for people with MND, carers, families and professionals, available in multiple formats and languages.
mndassociation.org/pro-info-finder
mndassociation.org/careinfofinder

Education

Our education programme aims to improve standards of care and quality of life. Opportunities include webinars and face to face equipment training.
mndassociation.org/education

MND Professionals' Community of Practice

A peer led group supporting cross disciplinary learning in MND care. Membership can contribute to CPD and offers access to networking and learning events.
mndassociation.org/cop

Local support

We offer online and local peer support, plus trained volunteers who provide practical help by phone, email or visits.
mndassociation.org/local-support

We fund and develop specialist care centres and networks across England, Wales and Northern Ireland, offering multidisciplinary care.
mndassociation.org/care-centres

Financial support

We offer a range of grants to support people living with MND, their families and unpaid carers. These are not in place of any statutory funding that should be available.
mndassociation.org/getting-support

MND register

The Register aims to collect information about everyone with MND in England, Wales and Northern Ireland to support care planning and research.
mndregister.ac.uk

Research into MND

We fund and promote research that leads to new understanding and treatments, and brings us closer to a cure for MND. We also produce information sheets on MND research for people with or affected by MND.
mndassociation.org/research

Acknowledgements

Thank you to the following people for their valuable contributions to this information sheet, and earlier versions:

Dan Harbour	Managing Director, Beacon Continuing Healthcare
Emma Wilkinson	Specialist Dietitian, NHS Lothian
Helen Copsey	MND Care and Research Network Coordinator, Norfolk and Norwich University Hospital, NHSFT
Kate Pottinger	Service Manager - Integrated Neighbourhoods, London Borough of Lewisham

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 **smartsurvey.co.uk/s/mndprofessionals** or email your comments to education@mndassociation.org.

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

How to order our publications

Our publications are free for anyone with or affected by MND or Kennedy's disease, including professionals. Health and social care professionals can also order items on behalf of someone with or affected by MND or Kennedy's disease.

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

Motor Neurone Disease Association
Francis Crick House
6 Summerhouse Road
Moulton Park
Northampton NN3 6BJ

Tel:	01604 250505
Email:	enquiries@mndassociation.org
Website:	mndassociation.org

Registered Charity no. 294354

Created: March 2026
Next review: March 2029
Version: 1

Every day we support people affected by Motor Neurone Disease.
Because with MND, every day matters.