Motor neurone disease: assessment and management

NICE guideline: short version

Draft for consultation, September, 2015

This guideline covers the assessment and management of motor neurone disease (MND).

Who is it for?

- Adults with MND, their families and carers.
- Healthcare professionals and social care practitioners caring for and supporting adults with MND.
- Commissioners and providers of MND health and social care services.

This version of the guideline contains the recommendations, context and recommendations for research. The Guideline Committee's discussion and the evidence reviews are in the full guideline.

Other information about how the guideline was developed is on the project page. This includes the scope, and details of the Committee and any declarations of interest.

This guideline amalgamates new guidance on the assessment and management of motor neurone disease with NICE guideline CG105 (published July 2010), and will replace NICE guideline CG105.

You are invited to comment on the new recommendations in this guideline.

These are marked as:

- **[new 2016]** if the evidence has been reviewed and the recommendation has been added.

You are also invited to comment on recommendations that NICE proposes to delete from the 2010 guideline.

We have not updated recommendations shaded in grey, and cannot accept comments on them. In some cases, we have made minor wording changes for clarification.

See How this guideline amalgamates with NICE guideline CG105 and Recommendations that have been deleted or changed for a full explanation of what is being updated.

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Contents

Recommendations

People have the right to be involved in discussions and make informed decisions about their care, as described in Your care.

Making decisions using NICE guidelines explains how we use words to show the strength of our recommendations, and has information about safeguarding, consent and prescribing medicines (including 'off-label' use).

1.1 Recognition and referral

1.1.1 Ensure that robust protocols and pathways are in place to:

- inform healthcare professionals about motor neurone disease (MND) and how it may present
- inform healthcare professionals in all settings about local referral arrangements
- ensure the continued and integrated care of people with MND across all care settings. **[new 2016]**

1.1.2 Be aware that MND causes progressive muscular weakness that may first present as isolated and unexplained symptoms. These symptoms may include:

- functional effects of muscle weakness, such as loss of dexterity, falls or trips
- speech or swallowing problems, or tongue fasciculations (this is known as bulbar presentation)
- muscle problems, such as weakness, wasting, twitching, cramps and stiffness
- breathing problems, such as shortness of breath on exertion or respiratory symptoms that are hard to explain
- effects of reduced respiratory function, such as excessive daytime sleepiness, fatigue, early morning headache or shortness of breath when lying down. **[new 2016]**

1.1.3 Be aware that MND may first present with cognitive changes, which may include:

- behavioural changes
- emotional lability (not related to dementia)
- frontotemporal dementia. **[new 2016]**

1.1.4 If you suspect MND, refer the person without delay and specify the possible diagnosis in the referral letter. Contact the consultant neurologist or specialist directly if you think the person needs to be seen urgently. **[new 2016]**

1.1.5 Provide support and information for people and their family members and/or carers (as appropriate) throughout the diagnostic process, particularly during periods of diagnostic uncertainty or delay. **[new 2016]**

1.2 Information and support at diagnosis

1.2.1 Information about the diagnosis, prognosis and management of MND should be given by a neurologist with up-to-date knowledge and experience of treating people with MND unless it is clinically necessary to give the diagnosis in an urgent situation. The neurologist should have knowledge and expertise in the following:

- Symptoms of MND.
- Types and possible causes of MND.
- Treatment options.
- How MND may progress (including cognitive and behavioural changes) and how progression may affect the treatments offered.
- Crisis prevention (for example, if there is an acute hospital admission or a breakdown in care arrangements).
- Opportunities for people with MND to be involved in research.
- Likely needs and concerns of people with MND and their family members and/or carers (as appropriate).

- Advance care planning. **[new 2016]**

1.2.2 Provide people with a single point of contact, and information about what to do if there are any concerns between assessments or appointments, during 'out-of-hours' or in an emergency, or if there is a problem with equipment. **[new 2016]**

1.2.3 When MND is suspected or confirmed, inform the person's GP without delay. **[new 2016]**

1.2.4 Ask people about how much information they wish to receive about MND, and about their preferences for involving their family members and/or carers (as appropriate). **[new 2016]**

1.2.5 Offer information and explanations about MND at diagnosis or when people ask for it. If the person agrees, share the information with their family members and/or carers (as appropriate). Information should be oral and written, and should include the following:

- What MND is.
- Types and possible causes.
- Likely symptoms and how they can be managed.
- How MND may progress.
- Treatment options.
- Where the person's appointments will take place.
- Which healthcare professionals and social care practitioners will undertake the person's care.
- Expected waiting times for consultations, investigations and treatments.
- Local services (including social care services) and how to get in touch with them.
- Local support groups, online forums and national charities, and how to get in touch with them.

- Legal rights, including social care support, employment rights and benefits.
- Requirements for disclosure, such as notifying the Driver and Vehicle Licensing Agency (DVLA).
- Opportunities for advance care planning. **[new 2016]**

1.2.6 Set aside enough time to discuss concerns, which may include the following:

- What will happen to me?
- Are there any treatments available?
- Is there a cure?
- How long will I live?
- What will the impact on my day-to-day life be?
- What will happen next with my healthcare?
- Will my children get MND?
- How do I tell my family and friends?
- How will I die? **[new 2016]**

1.2.7 If the person has any social care needs, refer them to social services for an assessment. Be aware that many people with MND may not have informal care available and may live alone or care for someone else. **[new 2016]**

1.2.8 Advise carers that they have a legal right to have a Carer's Assessment of their needs; support them with requesting this from their local authority. **[new 2016]**

1.3 Cognitive assessments

1.3.1 Be aware that people with MND and frontotemporal dementia may lack mental capacity. Care should be provided in line with the Mental Capacity Act 2005. **[new 2016]**

1.3.2 At diagnosis, and if there is concern about cognition and behaviour, explore any cognitive or behavioural changes with the person and

their family members and/or carers as appropriate. If needed, refer the person for a formal assessment in line with the NICE guideline on dementia. **[new 2016]**

1.3.3 Tailor all discussions to the person's needs, taking into account their communication ability, cognitive status and mental capacity. See the NICE guideline on patient experience in adult NHS services. **[new 2016]**

1.4 Prognostic factors

1.4.1 Estimate survival time on an individual basis. Take into account the following prognostic factors, which are associated with shorter survival if they are present at diagnosis:

- Speech and swallowing problems (bulbar presentation).
- Weight loss.
- Poor respiratory function.
- Older age.
- Lower Amyotrophic Lateral Sclerosis Functional Rating Scale (ALSFRS or ALSFRS-R) score.
- Shorter time from first developing symptoms to time of diagnosis. **[new 2016]**

1.5 Organisation of care

1.5.1 Provide coordinated care for people with MND, using a clinic-based, multidisciplinary team approach. **[new 2016]**

1.5.2 The multidisciplinary team should:

- include healthcare professionals and social care practitioners with expertise in MND, and staff who see people in their home
- ensure effective communication between all healthcare professionals and social care practitioners involved in the person's care and their family members and/or carers (as appropriate)

- carry out regular, coordinated assessments at the multidisciplinary team clinic (usually every 2–3 months) to assess people's symptoms and needs. **[new 2016]**

1.5.3 The multidisciplinary team should assess the following:

- Weight, diet, nutritional intake, feeding and swallowing (see recommendations 1.10.1–1.10.10).
- Muscle problems, such as weakness, stiffness, cramps (see recommendations 1.8.1–1.8.9).
- Physical function, including mobility and activities of daily living (see recommendations 1.9.1–1.9.8).
- Saliva problems, such as drooling of saliva (sialorrhoea) and thick, tenacious saliva (see recommendations 1.8.10–1.8.15).
- Speech and communication (see recommendations 1.11.1–1.11.6).
- Cough effectiveness (see recommendations 1.12.1–1.12.4).
- Respiratory function (see section 1.13).
- Pain and other symptoms, such as constipation.
- Cognition and behaviour (see recommendations 1.3.1–1.3.3).
- Psychological support needs (see recommendations 1.6.1–1.6.4).
- Social care needs (see recommendations 1.6.5–1.6.6).
- Information and support needs for the person and their family members and/or carers (as appropriate). **[new 2016]**

1.5.4 The core multidisciplinary team should consist of healthcare professionals and other professionals with expertise in MND, and should include the following:

- Neurologist.
- Specialist nurse.
- Dietitian.
- Physiotherapist.
- Occupational therapist.

- Respiratory physiologist or a healthcare professional who can assess respiratory function.
- Speech and language therapist. **[new 2016]**

1.5.5 The multidisciplinary team should have access to the following services:

- Clinical psychology and/or neuropsychology.
- Social care.
- Counselling.
- Respiratory medicine.
- Specialist palliative care.
- Gastroenterology. **[new 2016]**

1.5.6 Tailor the frequency of the multidisciplinary team assessments to the person's symptoms and needs, with more or less frequent assessments as needed. **[new 2016]**

1.5.7 Ensure arrangements are in place to trigger an earlier multidisciplinary team assessment if there is a significant change in symptoms identified by family members and/or carers (as appropriate), or healthcare professionals. **[new 2016]**

1.5.8 Tailor the multidisciplinary team assessment to the person's needs, for example, adjust the format if the person has cognitive or behaviour changes or difficulties with communication. **[new 2016]**

1.5.9 Inform all healthcare professionals and social care practitioners involved in the person's care about key decisions reached with the person and their family members and/or carers (as appropriate). **[new 2016]**

1.5.10 Ensure that all healthcare professionals and social care practitioners involved in the person's care are aware that MND symptoms may get worse quickly, and that people with MND will need repeated assessment. Priority should be given to ensuring

continuity of care and avoidance of untimely case closure. **[new 2016]**

1.6 Psychological and social care support

1.6.1 During multidisciplinary team assessments and other appointments, discuss the psychological and emotional impact of MND with the person and ask whether they have any psychological or support care needs. Topics to discuss may include the following:

- Their understanding of MND and how it affects daily living.
- Accepting and coping with the diagnosis and prognosis, including concerns and fears about dying.
- Adjusting to changes in their life and their perception of self.
- Changes in relationships, familial roles and family dynamics.
- Sexuality and intimacy.
- Concerns about their family members and/or carers.
- Decision-making. **[new 2016]**

1.6.2 Offer the person information about sources of emotional and psychological support, including support groups and online forums. If needed, refer them to psychological services for a specialist assessment and support. **[new 2016]**

1.6.3 During multidisciplinary team assessments and other appointments, discuss the psychological and emotional impact of MND with family members and/or carers (as appropriate), and ask whether they have any psychological or social care support needs. Topics to discuss may include the following:

- Their understanding of MND and how it affects daily living.
- Accepting and coping with the diagnosis and prognosis, including concerns and fears about the person with MND dying.
- Adjusting to changes in their life.
- Changes in relationships, familial roles and family dynamics, including their change to a carer role (if appropriate).

- Sexuality and intimacy.
- Involvement in decision-making.
- Impact on other family members and/or carers.
- Ability and willingness to provide personal care and operate equipment. **[new 2016]**

1.6.4 Offer family members and/or carers (as appropriate) information about respite care and sources of emotional and psychological support, including support groups and online forums. **[new 2016]**

1.6.5 A social care practitioner with knowledge of MND or rapidly progressive complex disabilities should discuss the person's needs, and preferences for social care, and provide information and support for them to access the following:

- Personal care, ensuring there is continuity of care with familiar workers, so that wherever possible, personal care and support is carried out by workers known to the person and their family members and/or carers (as appropriate).
- Equipment and practical support (see recommendations 1.9.1–1.9.8).
- Financial support and advice (for example, money management, how to access carers' and disability benefits and grants, continuing healthcare funding and funeral expenses).
- Support to engage in social activities and hobbies, such as access to social media and physical access to activities outside their home.
- Respite care. **[new 2016]**

1.6.6 Be aware that as MND progresses, people may develop communication problems and have difficulty accessing support or services. For example, they may be unable to access a call centre. Ensure that they are given different ways of getting in touch with support or services, and a designated contact if possible. **[new 2016]**

1.7 Planning for end of life

1.7.1 Offer the person with MND the opportunity to discuss their preferences and concerns about care at the end of life at trigger points such as: at diagnosis, if there is a significant change in respiratory function, or if interventions such as gastrostomy or non-invasive ventilation are needed. Be sensitive about the timing of discussions and take into account the person's current communication ability, cognitive status and mental capacity. **[new 2016]**

1.7.2 Think about discussing advance care planning with people at an earlier opportunity if you expect their communication ability, cognitive status or mental capacity to get worse. **[new 2016]**

1.7.3 Consider an early referral to a specialist palliative care team for people with significant or complex needs, such as psychological or social distress or rapidly progressing symptoms. **[new 2016]**

1.7.4 Provide support and advice on advance care planning for end of life to the person with MND and their family members and/or carers (as appropriate). The discussion should include:

- What could happen at end of life, for example how death may occur.
- Providing anticipatory medicines in the home.
- Advance care planning, including Advanced Decisions to Refuse Treatment (ADRT) and Do Not Attempt Resuscitation (DNACPR) orders, and Lasting Power of Attorney.
- Areas that people might wish to plan for, such as:
  - what they want to happen (for example preferred place of death)
  - what they do not want to happen (for example being admitted to hospital)
  - who will represent their decisions, if necessary

- what should happen if they develop an intercurrent illness. **[new 2016]**

1.7.5 Offer people the opportunity to talk about ADRT, DNACPR and Lasting Power of Attorney when interventions such as gastrostomy and non-invasive ventilation are planned. **[new 2016]**

1.7.6 Provide additional support as the end of life approaches, for example, additional social or nursing care to enable informal carers and family to reduce their carer responsibilities and spend time with the person with MND. **[new 2016]**

1.7.7 Towards the end of life, ensure there is access to the following:

- An appropriate method of communication, such as an alternative and augmentative communication (AAC) system.
- Holistic support.
- Specialist palliative care.
- Equipment, if needed, such as syringe drivers, suction machines, riser–recliner chair, hospital bed, commode, hoist.
- Anticipatory medicines including opioids and benzodiazepines to treat breathlessness, and anticholinergic medicines to treat problematic saliva and respiratory secretions. **[new 2016]**

1.7.8 Offer bereavement support to family members and/or carers (as appropriate). **[new 2016]**

1.8 Managing symptoms

Pharmacological treatments for muscle problems

1.8.1 Discuss the available treatment options for muscle problems. Take into account the person's needs and preferences, and whether they have any difficulties taking medicine (for example, if they have problems swallowing). **[new 2016]**

1.8.2 Consider quinine[1] as first-line treatment for muscle cramps in people with MND. If quinine is not effective, not tolerated or contraindicated, consider baclofen[1] instead as second-line treatment. If baclofen is not effective, not tolerated or contraindicated, consider tizanidine[1], dantrolene[1] or gabapentin[1]. **[new 2016]**

1.8.3 Consider baclofen, tizanidine, dantrolene[1] or gabapentin[1] to treat muscle stiffness, spasticity or increased tone in people with MND. If these treatments are not effective, not tolerated or contraindicated, consider referral to a specialist service for treatment of severe spasticity. **[new 2016]**

1.8.4 Review the treatments for muscle problems during multidisciplinary team assessments, ask about how the person is finding the treatment, whether it is working and whether they have any adverse side effects. **[new 2016]**

Exercise programmes

1.8.5 Consider an exercise programme for people with MND to:

- maintain joint range of movement
- prevent contractures
- reduce stiffness and discomfort
- optimise function and quality of life. **[new 2016]**

1.8.6 Choose a programme that is appropriate to the person's level of function and tailored to their needs, abilities and preferences. Take into account factors such as postural needs and fatigue. The programme might be a resistance programme, an active-assisted programme or a passive programme. **[new 2016]**

[1] At the time of consultation (September 2015), these medicines did not have a UK marketing authorisation for this indication. The prescriber should follow relevant professional guidance, taking full responsibility for the decision. Informed consent should be obtained and documented. See the General Medical Council's Prescribing guidance: prescribing unlicensed medicines for further information.

1.8.7 Check that family members and/or carers (as appropriate) are willing and able to help with exercise programmes. **[new 2016]**

1.8.8 Give advice to the person and their family members and/or carers (as appropriate) about safe manual handling. **[new 2016]**

1.8.9 If a person needs orthoses to help with muscle problems, they should be referred without delay, and the orthoses should be provided without delay. **[new 2016]**

Saliva problems

1.8.10 If a person with MND has problems with saliva, assess the volume, colour and viscosity of the saliva and the person's respiratory function, swallowing, diet, posture and oral care. **[new 2016]**

1.8.11 If a person with MND has problems with drooling of saliva (sialorrhoea), provide advice on swallowing, diet, posture, positioning, oral care and suctioning. **[new 2016]**

1.8.12 Consider a trial of anticholinergic medicine[2] as the first-line treatment for sialorrhoea in people with MND. **[new 2016]**

1.8.13 Consider glycopyrrolate[2] as the first-line treatment for sialorrhoea in people with MND who have cognitive impairment, because it has fewer central nervous system side effects. **[new 2016]**

1.8.14 If first-line treatment for sialorrhoea is not effective, not tolerated or contraindicated, consider referral to a specialist service for Botulinum toxin A. **[new 2016]**

1.8.15 If a person with MND has thick, tenacious saliva:

- review all current medicines, especially any treatments for sialorrhoea

[2] At the time of consultation (September 2015), these medicines did not have a UK marketing authorisation for this indication. The prescriber should follow relevant professional guidance, taking full responsibility for the decision. Informed consent should be obtained and documented. See the General Medical Council's Prescribing guidance: prescribing unlicensed medicines for further information.

- provide advice on swallowing, diet, posture, positioning, oral care, suctioning and hydration
- consider treatment with humidification, nebulisers and carbocisteine. **[new 2016]**

1.9 Equipment and adaptations to aid activities of daily living and mobility

1.9.1 Healthcare professionals and social care practitioners, which will include physiotherapists and occupational therapists, should assess the person's daily living needs, taking into account the following:

- Activities of daily living, including personal care, dressing and bathing, and eating and drinking.
- Mobility and avoiding falls and problems from loss of dexterity.
- The home environment and the need for adaptations.
- The need for assistive technology, such as environmental control systems. **[new 2016]**

1.9.2 Provide appropriate equipment and adaptations without delay to maximise people's participation in activities of daily living and maintain their quality of life. **[new 2016]**

1.9.3 Refer people to specialist services without delay if assistive technology such as environmental control systems is needed. People should be assessed and assistive technology provided without delay. **[new 2016]**

1.9.4 Refer people to wheelchair services without delay if needed. Wheelchair needs should be assessed and a wheelchair provided without delay. **[new 2016]**

1.9.5 Ensure that equipment, adaptations, daily living aids, assistive technology and wheelchairs meet the changing needs of the

person and their family and/or carers (as appropriate) to maximise mobility and participation in activities of daily living. **[new 2016]**

1.9.6 Ensure regular, ongoing monitoring of the person with MND's mobility and daily life needs and abilities as their disease progresses and regularly review their ability to use equipment and to adapt equipment as necessary. **[new 2016]**

1.9.7 Healthcare professionals, social care practitioners and other services providing equipment should liaise to ensure that all equipment provided can be integrated, for example, integrating AAC aids and devices and environmental control systems with wheelchairs. **[new 2016]**

1.9.8 Enable speedy access and assessment for funding for home adaptation. If the person is not eligible for funding, continue to offer information and support in arranging home environment adaptations. **[new 2016]**

1.10 Nutrition and gastrostomy

1.10.1 At diagnosis and at multidisciplinary team assessments, or if there are any concerns about weight, nutrition or swallowing, assess the person's weight, diet, nutritional intake, oral health, feeding and swallowing, and offer support, advice and interventions as needed (see recommendations 1.10.2–1.10.10). See the NICE guideline on nutrition support in adults. **[new 2016]**

1.10.2 Assess the person's diet and nutritional intake by taking into account:

- intake versus nutritional needs of both solids and liquids
- nutritional supplements, if needed
- appetite
- gastrointestinal symptoms, such as nausea or constipation

- causes of reduced oral intake (for example, swallowing difficulties, limb weakness or the possibility of low mood or depression causing loss of appetite). **[new 2016]**

1.10.3 Assess the person's ability to feed by taking into account:

- the need for feeding aids and altered utensils to help them take food from the plate to their mouth
- the need for help with food preparation
- advice and aids for positioning, seating and posture while feeding
- dealing with social situations (for example, eating out). **[new 2016]**

1.10.4 Arrange for a formal swallowing assessment if swallowing problems are suspected. **[new 2016]**

1.10.5 Assess factors that may contribute to problems with swallowing, such as:

- positioning
- seating
- the need to modify food and drink consistency and palatability
- respiratory symptoms and risk of aspiration and/or choking
- fear of choking and psychological considerations (for example wanting to eat normally in social situations). **[new 2016]**

1.10.6 Pay particular attention to the nutritional needs of people with MND who have frontotemporal dementia and who lack mental capacity. The multidisciplinary team assessment should include the support they need from carers, and their ability to understand the risks of swallowing difficulties. **[new 2016]**

1.10.7 Discuss gastrostomy at an early stage, and at regular intervals as MND progresses, taking into account the person's preferences and issues, such as ability to swallow, weight loss, respiratory function,

effort of feeding and risk of choking. Be aware that some people will not wish to have a gastrostomy. **[new 2016]**

1.10.8 Explain the benefits of early placement of a gastrostomy, and the possible risks of a late gastrostomy (for example, low critical body mass, respiratory complications, risk of dehydration, different methods of insertion, and a higher risk of mortality and procedural complications). **[new 2016]**

1.10.9 If a person is referred for a gastrostomy, it should take place without unnecessary delay. **[new 2016]**

1.10.10 Before a decision is made on the use of gastrostomy for a person with MND who has frontotemporal dementia, the neurologist from the multidisciplinary team should assess the following:

- The person's ability to make decisions and to give consent[3].
- The severity of frontotemporal dementia and cognitive problems.
- Whether the person is likely to accept and cope with treatment.

Discuss with the person's family members and/or carers (as appropriate; with the person's consent if they have the ability to give it). **[new 2016]**

1.11 Communication

1.11.1 When assessing speech and communication needs during multidisciplinary team assessments and other appointments, discuss face-to-face and remote communication, for example using the telephone, email, the internet and social media. Ensure that the assessment and review is carried out by a speech and language therapist. **[new 2016]**

1.11.2 Provide AAC equipment that meets the needs of the person without delay to maximise participation in activities of daily living and maintain quality of life. The use of both low level technologies, for

[3] See Mental Capacity Act 2005.

example, alphabet, word or picture boards and high level technologies, for example PC or tablet-based voice output communication aids may be helpful. Review the person's communication needs during multidisciplinary team assessments. **[new 2016]**

1.11.3 Liaise with, or refer the person with MND to, a specialised NHS AAC hub if complex high technology AAC equipment (for example, eye gaze access) is needed. **[new 2016]**

1.11.4 Involve other healthcare professionals, such as occupational therapists, to ensure that AAC equipment is integrated with other assistive technologies, such as environmental control systems. **[new 2016]**

1.11.5 Ensure regular, ongoing monitoring of the person's communication needs and abilities as MND progresses, and review their ability to use AAC equipment. Reassess and liaise with a specialised NHS AAC hub if needed. **[new 2016]**

1.11.6 Provide ongoing support and training for the person with MND, and their family members and/or carers (as appropriate), in using AAC equipment and other communication strategies. **[new 2016]**

1.12 Cough effectiveness

1.12.1 Offer cough augmentation techniques to people with MND who cannot cough effectively. **[new 2016]**

1.12.2 Consider breath stacking as the first-line treatment for people with MND who have an ineffective cough. **[new 2016]**

1.12.3 For patients with bulbar dysfunction, or whose cough is ineffective with unassisted breath stacking, consider assisted breath stacking (for example, using a lung volume recruitment bag). **[new 2016]**

1.12.4 Consider a mechanical cough assist device if assisted breath stacking is not effective, and/or during a respiratory tract infection. **[new 2016]**

1.13 Respiratory impairment

1.13.1 Assess and treat people with MND and worsening respiratory impairment for reversible causes (for example, respiratory tract infections or secretion problems) before considering other treatments. **[new 2016]**

1.13.2 Offer non-invasive ventilation as treatment for people with respiratory impairment (see section 1.14). **[new 2016]**

1.13.3 Consider urgent introduction of non-invasive ventilation for people with MND who develop worsening respiratory impairment and are not already using non-invasive ventilation. **[new 2016]**

1.13.4 Decisions to offer non-invasive ventilation should be made by the multidisciplinary team (see recommendations 1.5.1–1.5.5). **[new 2016]**

1.13.5 Consider opioids[4] as an option to relieve symptoms of breathlessness. Take into account the route of administration and acquisition cost of medicines. **[new 2016]**

1.13.6 Consider benzodiazepines to manage breathlessness that is exacerbated by anxiety. Take into account the route of administration and acquisition cost of medicines. **[new 2016]**

[4] At the time of consultation (September 2015), these medicines did not have a UK marketing authorisation for this indication. The prescriber should follow relevant professional guidance, taking full responsibility for the decision. Informed consent should be obtained and documented. See the General Medical Council's Prescribing guidance: prescribing unlicensed medicines for further information.

1.14 Non-invasive ventilation

Information and support about non-invasive ventilation

1.14.1 Offer to discuss the possible use of non-invasive ventilation with the person and (if the person agrees) their family and carers, at an appropriate time and in a sensitive manner. This may be at one or more of the following times:

- soon after MND is first diagnosed
- when monitoring respiratory function
- when respiratory function deteriorates
- if the person asks for information. **[2010]**

1.14.2 Discussions about non-invasive ventilation should be appropriate to the stage of the person's illness, carried out in a sensitive manner and include information on:

- the possible symptoms and signs of respiratory impairment (see table 1)
- the purpose, nature and timing of respiratory function tests, and explanations of the test results
- how non-invasive ventilation (as a treatment option) can improve symptoms associated with respiratory impairment and can be life prolonging, but does not stop progression of the underlying disease. **[2010, amended 2016]**

1.14.3 When discussing non-invasive ventilation, explain the different ways that people can manage their breathlessness symptoms. This should include:

- non-invasive ventilation, and its advantages and disadvantages
- using non-invasive ventilation at different points in the course of the person's lifetime
- the possibility of the person becoming dependent on non-invasive ventilation

- options for treating any infections
- support and information on how to recognise and cope with a distressing situation
- the role of medications
- psychological techniques and support. **[new 2016]**

1.14.4 Check that the person thinking about non-invasive ventilation:

- understands what non-invasive ventilation is and what it can achieve
- recognises the need for regular review
- has enough information about non-invasive ventilation and other options for breathing problems to make decisions about how and when to use it. **[new 2016]**

1.14.5 Explain that non-invasive ventilation can be stopped at any time. Reassure people that they can ask for help and advice if they need it, especially if they are dependent on non-invasive ventilation for 24 hours a day, or become distressed when attempting to stop it. **[new 2016]**

1.14.6 Ensure that families and carers:

- have an initial assessment if the person they care for decides to use non-invasive ventilation, which should include:
  - their ability and willingness to assist in providing non-invasive ventilation
  - their training needs
- have the opportunity to discuss any concerns they may have with members of the multidisciplinary team and/or other healthcare professionals. **[2010]**

Identification and assessment of respiratory impairment

Symptoms and signs

1.14.7 Monitor the symptoms and signs listed in table 1 to detect potential respiratory impairment. **[2010, amended 2016]**

Table 1 Symptoms and signs of potential respiratory impairment

Symptoms	Signs
Breathlessness	Increased respiratory rate
Orthopnoea	Shallow breathing
Recurrent chest infections	Weak cough[1]
Disturbed sleep	Weak sniff
Non-refreshing sleep	Abdominal paradox (inward movement of the abdomen during inspiration)
Nightmares	Use of accessory muscles of respiration
Daytime sleepiness	Reduced chest expansion on maximal inspiration
Poor concentration and/or memory	
Confusion	
Hallucinations	
Morning headaches	
Fatigue	
Poor appetite	

[1] Weak cough could be assessed by measuring cough peak flow.

Respiratory function tests

1.14.8 As part of the initial assessment to diagnose MND, or soon after diagnosis, a healthcare professional from the multidisciplinary team who has appropriate competencies should perform the following tests (or arrange for them to be performed) to establish the person's baseline respiratory function:

- oxygen saturation measured by pulse oximetry (SpO_2):
  - this should be a single measurement of SpO_2 with the person at rest and breathing room air
  - if it is not possible to perform pulse oximetry locally, refer the person to a specialist respiratory service.

Then one or both of the following:

- forced vital capacity (FVC) or vital capacity (VC)[5]
- sniff nasal inspiratory pressure (SNIP) and/or maximal inspiratory pressure (MIP). **[2010]**

1.14.9 If the person has severe bulbar impairment or severe cognitive problems that may be related to respiratory impairment:

- ensure that SpO_2 is measured (at rest and breathing room air)
- do not perform the other respiratory function tests (FVC, VC, SNIP and MIP) if interfaces are not suitable for the person. **[2010]**

1.14.10 A healthcare professional with appropriate competencies should perform the respiratory function tests every 2–3 months, although tests may be performed more or less often depending on:

- whether there are any symptoms and signs of respiratory impairment (see table 1)
- the rate of progression of MND
- the person's preference and circumstances. **[2010, amended 2016]**

1.14.11 Perform arterial or capillary blood gas analysis if the person's SpO_2 (measured at rest and breathing room air):

- is less than or equal to 92% if they have known lung disease
- is less than or equal to 94% if they do not have lung disease.

If it is not possible to perform arterial or capillary blood gas analysis locally, refer the person to a specialist respiratory service. **[2010]**

1.14.12 If the person's SpO_2 (measured at rest and breathing room air) is greater than 94%, or 92% for those with lung disease, but they have sleep-related respiratory symptoms:

[5] The difference between the measurement of vital capacity and forced vital capacity is very subtle and so either can be used.

- consider referring them to a specialist respiratory service for nocturnal (overnight) oximetry and/or a limited sleep study **and**
- discuss both the impact of respiratory impairment and treatment options with the patient and (if the person agrees) their family and carers. **[2010]**

1.14.13 If the person's arterial partial pressure of carbon dioxide ($PaCO_2$) is greater than 6 kPa:

- refer them urgently to a specialist respiratory service (to be seen within 1 week) and
- explain the reasons for and implications of the urgent referral to the person and (if the person agrees) their family and carers. **[2010]**

1.14.14 If the person's $PaCO_2$ is less than or equal to 6 kPa but they have any symptoms or signs of respiratory impairment, particularly orthopnoea (see recommendation 1.14.15):

- refer them to a specialist respiratory service for nocturnal (overnight) oximetry and/or a limited sleep study **and**
- discuss both the impact of respiratory impairment and treatment options with the person and (if the person agrees) their family and/or carers (as appropriate). **[2010]**

1.14.15 If any of the results listed in table 2 is obtained, discuss with the person and (if appropriate) their family and carers:

- their respiratory impairment
- their treatment options
- possible referral to a specialist respiratory service for further assessment based on discussion with the person, and their wishes. **[2010, amended 2016]**

Table 2 Results of respiratory function tests

Forced vital capacity (FVC) or vital capacity (VC)	Sniff nasal inspiratory pressure (SNIP) and/or maximal inspiratory pressure (MIP) (if both tests are performed, base the assessment on the better respiratory function reading)
FVC or VC less than 50% of predicted value FVC or VC less than 80% of predicted value plus any symptoms or signs of respiratory impairment (see recommendation 1.14.7), particularly orthopnoea	SNIP or MIP less than 40 cmH_2O SNIP or MIP less than 65 cmH_2O for men or 55 cmH2O for women plus any symptoms or signs of respiratory impairment (see recommendation 1.14.7), particularly orthopnoea Repeated regular tests show a rate of decrease of SNIP or MIP of more than 10 cm H_2O per 3 months

People with a diagnosis of frontotemporal dementia

1.14.16 Base decisions on respiratory function tests for a person with a diagnosis of frontotemporal dementia on considerations specific to their needs and circumstances, such as:

- their ability to give consent[6]
- their understanding of the tests
- their tolerance of the tests and willingness to undertake them
- the impact on their family and carers
- whether they are capable of receiving non-invasive ventilation.

[2010, amended 2016]

Non-invasive ventilation for treatment of respiratory impairment in people with MND

1.14.17 Offer a trial of non-invasive ventilation if the person's symptoms and signs and the results of the respiratory function tests indicate that the person is likely to benefit from the treatment. **[2010, amended 2016]**

[6] See Mental Capacity Act 2005.

1.14.18 Consider a trial of non-invasive ventilation for a person who has severe bulbar impairment or severe cognitive problems that may be related to respiratory impairment only if they may benefit from an improvement in sleep-related symptoms or correction of hypoventilation. **[2010, amended 2016]**

1.14.19 Before starting non-invasive ventilation, the multidisciplinary team should carry out and coordinate a patient-centred risk assessment, after discussion with the person and their family and carers. This should consider:

- the most appropriate type of non-invasive ventilator and interfaces, based on the person's needs and lifestyle factors
- the person's tolerance of the treatment
- the risk, and possible consequences, of ventilator failure
- the power supply required, including battery back-up
- how easily the person can get to hospital
- risks associated with travelling away from home (especially abroad)
- whether a humidifier is required
- issues relating to secretion management
- the availability of carers. **[2010]**

1.14.20 Before starting non-invasive ventilation, the multidisciplinary team should prepare a comprehensive care plan, after discussion with the person and their family and carers (who should be offered a copy of the plan). This should cover:

- long-term support provided by the multidisciplinary team
- the initial frequency of respiratory function tests and monitoring of respiratory impairment
- the frequency of clinical reviews of symptomatic and physiological changes
- the provision of carers

- arrangements for device maintenance and 24-hour emergency clinical and technical support
- secretion management and respiratory physiotherapy assessment, including cough-assist therapy (if required)
- training in and support for the use of non-invasive ventilation for the person and their family and carers
- regular opportunities to discuss the person's wishes in relation to continuing or withdrawing non-invasive ventilation. **[2010, amended 2016]**

1.14.21 When starting non-invasive ventilation:

- perform initial acclimatisation during the day when the person is awake
- usually start regular treatment at night, before and during sleep
- gradually build up the person's hours of use as necessary. **[2010]**

1.14.22 Continue non-invasive ventilation if the clinical reviews show:

- symptomatic and/or physiological improvements for a person without severe bulbar impairment and without severe cognitive problems
- an improvement in sleep-related symptoms for a person with severe bulbar impairment or with severe cognitive problems that may be related to respiratory impairment. **[2010]**

1.14.23 Provide the person and their family and/or carers (as appropriate) with support and assistance to manage non-invasive ventilation. This should include:

- training on using non-invasive ventilation and ventilator interfaces, for example:
  - emergency procedures

  - night-time assistance if the person is unable to use the equipment independently (for example, emergency removal or replacement of interfaces)
  - how to use the equipment with a wheelchair or other mobility aids if required
  - what to do if the equipment fails
- assistance with secretion management
- information on general palliative strategies
- an offer of ongoing emotional and psychological support for the person and their family and carers. **[2010, amended 2016]**

1.14.24 Discuss all decisions to continue or withdraw non-invasive ventilation with the person and (if the person agrees) their family and carers. **[2010]**

1.14.25 Before a decision is made on the use of non-invasive ventilation for a person with a diagnosis of frontotemporal dementia, the multidisciplinary team should carry out an assessment that includes:

- the person's capacity to make decisions and to give consent[7]
- the severity of dementia and cognitive problems
- whether the person is likely to accept treatment
- whether the person is likely to achieve improvements in sleep-related symptoms and/or behavioural improvements
- a discussion with the person's family and/or carers (with the person's consent if they have the capacity to give it). **[2010, amended 2016]**

1.14.26 Consider prescribing medicines to help ease breathlessness that people using non-invasive ventilation can take on an 'as-needed' basis at home, for example, opioids or benzodiazepines. **[new 2016]**

[7] See Mental Capacity Act 2005.

1.14.27 Inform services that may see the person in crisis situations, such as their GP and services that provide emergency or urgent care, that the person is using non-invasive ventilation. **[new 2016]**

Stopping non-invasive ventilation

1.14.28 The healthcare professionals responsible for starting non-invasive ventilation treatment in people with MND should ensure that support is available for other healthcare professionals who may be involved if there is a plan to stop non-invasive ventilation, including the legal and ethical implications. **[new 2016]**

1.14.29 If a person on continuous non-invasive ventilation wishes to stop treatment, ensure that they have support from healthcare professionals with knowledge and expertise of:

- stopping non-invasive ventilation
- the ventilator machine
- palliative medication (see the NICE guideline on care of the dying adult [hyperlink to be added at publication])
- supporting the person, family members and/or carers (as appropriate)
- supporting other healthcare professionals involved with the person's care
- legal and ethical frameworks and responsibilities. **[new 2016]**

1.14.30 If a person on continuous non-invasive ventilation wishes to stop treatment, seek advice from healthcare professionals who have knowledge and experience of stopping non-invasive ventilation. **[new 2016]**

1.14.31 Healthcare professionals involved in stopping non-invasive ventilation should have up-to-date knowledge of the law regarding The Mental Capacity Act, DNACPR, ADRT and Lasting Power of Attorney. **[new 2016]**

To find out what NICE has said on topics related to this guideline, see our web page on motor neurone disease.

Implementation: getting started

This section will be completed in the final guideline using information provided by stakeholders during consultation.

To help us complete this section, please use the stakeholder comments form give us your views on these questions:

1. Which areas will have the biggest impact on practice and be challenging to implement? Please say for whom and why.

2. What would help users overcome any challenges? (For example, existing practical resources or national initiatives, or examples of good practice.)

Context

Motor neurone disease (MND) is a neurodegenerative condition affecting the brain and spinal cord. MND is characterised by the degeneration of primarily motor neurones, leading to muscle weakness.

There are several forms of MND:

- Amyotrophic lateral sclerosis (ALS) is characterised by muscle weakness, wasting and stiffness and affects about 66% of people with MND.
- Progressive bulbar palsy, in which the nerves first affected are those involved in speech and swallowing, and occurs in about 25% of people.
- Progressive muscular atrophy, which leads to muscle weakness and wasting of either arms or legs first, and affects about 10% of people with MND.

- Primary lateral sclerosis, which leads to muscle stiffness and has a longer prognosis of up to 10–15 years, and has no clear prevalence but is very rare.

The initial stages of MND may be one of these forms, but as the disease progresses the pattern of symptoms and signs become similar, with increasing muscle weakness in the person's arms and legs, problems swallowing and communicating and weakness of the muscles used for breathing, which ultimately leads to death.

Every person with MND has an individual progression of the disease. About 10–15% of people with MND will show signs of frontotemporal dementia, which causes cognitive dysfunction and issues in decision-making. A further 50% of people with MND may show signs of mild cognitive change, which may affect their ability to make decisions and plan ahead. Rarely, people are found to have MND after having developed breathing problems, due to weakness of the breathing muscles.

MND mainly affects people aged 50 to 65 years. Most people die within 2–3 years of developing symptoms, but 25% are alive at 5 years and 10% at 10 years. There are about 5000 people with MND in the UK and about 1100 people are diagnosed every year. The cause is not known although there is increasing evidence of a genetic basis. About 5–10% of people with MND have a family history of the disease and several abnormal genes have been identified. As there is no cure for MND, care focuses on maintaining functional ability and enabling people with MND and their family members to live as full a life as possible. Early diagnosis, without delay after investigation, may be helpful, as it allows medication and the provision of aids, as well as communication about the disease and advance care planning to be undertaken appropriately.

Care of people with MND varies across England and Wales, with MND care centres providing co-ordinated multidisciplinary care. However, some people with MND are left isolated and their care is less than ideal. This guideline aims to consider the clinical and cost-effectiveness evidence for the care of people

with MND from the time of diagnosis, including communication of the diagnosis, monitoring of disease progression, management of symptoms (in particular swallowing, breathing, muscle weakness and secretions), ongoing support and services available, mobility, emotional and psychological changes, and the preparation for end of life care. Particular emphasis is placed on the importance of a multidisciplinary team approach to the care and management of people with MND.

Recommendations for research

The Guideline Committee has made the following recommendations for research. The Committee's full set of research recommendations is detailed in the full guideline.

1 Cognitive assessment

What is the impact of assessing for cognitive and behaviour change in people with MND on clinical practice, the person and their family and carers? Does repeated assessment provide more benefit than assessment at a single point at diagnosis?

Why this is important

Clinic-based and population-based studies demonstrate that up to 15% of people with MND have frontotemporal dementia. A further third of people with MND have changes in behaviour and cognition. These impairments are present at diagnosis. Their course during the disease has shown varying patterns between studies although several studies have shown that cognitive and behavioural impairments predict poorer survival and increased carer burden. A randomised controlled trial is needed to assess whether formal assessment at diagnosis and/or repeated assessment improves clinical practice, subsequent care of the person and quality of life for the person, their family and carers.

2 Prognostic tools

Is the ALS Prognostic Index an accurate predictor of survival in people with MND under NHS care in England and/or Wales?

Why this is important

Accurate predictions of survival in people with MND would be of great use to clinicians and to the person with MND, their family and carers. Accurate predictions would enable people with MND to be clearer about their prognosis, make plans for the rest of their life and have a well-prepared and dignified transition into the end of life phase. Family members would similarly benefit in being more aware of the likely progression and prepare themselves for the death of their loved one.

Accurate predictions of survival would enable professionals to create and deliver more effective management and care plans and access services when it is most appropriate, for example specialist palliative care.

The ALS Prognostic Index (ALS-PI) was developed in a cohort of people with ALS in the Republic of Ireland and externally validated in a cohort in Italy. However, it has not been validated in people with ALS, primary lateral sclerosis or progressive muscular atrophy in the NHS in England or Wales. The tool needs to be validated in a UK population using a simplified measure of executive function.

3 Saliva

How is excessive drooling of saliva (sialorrhoea) managed in people with MND?

Why this is important

Sialorrhoea affects up to 50% of people with MND and in 42% of these individuals the symptom is poorly controlled. There is no evidence base for clinicians to make decisions with regards to the treatment options available. Anticholinergics are used first-line but there is no evidence to inform which anticholinergic and at what dose. Botulinum toxin is used second- or third-line

although there is little evidence to guide dosing, which salivary glands to inject and which type of botulinum toxin to use. Currently there is no baseline information about how specialists are using these treatments and this information is required to inform comparative studies.

4 Nutrition

Does a high calorific diet prolong survival of people with MND if initiated following diagnosis or following initiation of feeding using a gastrostomy?

Why this is important

There is little specific guidance on the optimal calorie intake for people with MND. There is growing evidence that people with MND have a hypercatabolic state and have high energy requirements. A large cohort study in the UK has demonstrated that nearly half of people continue to lose weight following gastrostomy and most show no improvement in their weight. A small study has demonstrated that high fat and high carbohydrate feeding may prolong survival in gastrostomy-fed people. A larger randomised trial is needed to inform clinical practice.

5 Augmentative and alternative communication

What is the current pattern of provision and use of augmentative and alternative communication (AAC) by people with MND in England?

Why this is important

Appropriate AAC equipment can have a significant effect on quality of life for people with MND. While the NHS has a responsibility to provide equipment and ongoing support in its use, there are no reliable data on the types of equipment found most useful at different stages of the disease process, or the number of people with MND who may benefit from AAC. A prospective census study of people with MND presenting with early onset of speech problems is needed to establish the current baseline provision and needs of this population and how best to utilise AAC equipment. The programme will begin

with the collection and analysis of basic data. It will then progress to patient-related outcomes.

How this guideline amalgamates with NICE guideline CG105

This guidance amalgamates new guidance on the assessment and management of motor neurone disease with NICE guideline CG105 (published July 2010), and will replace NICE guideline CG105.

New recommendations have been added for recognition and referral, information and support at diagnosis, cognitive assessments, prognostic factors, psychosocial and social support, organisation of care, planning for end of life care and managing symptoms for people with motor neurone disease.

These are marked as:

- **[new 2016]** if the evidence has been reviewed and the recommendation has been added.

NICE proposes to delete some recommendations from the 2010 guideline, because either the evidence has been reviewed and the recommendations have been updated, or NICE has updated other relevant guidance and has replaced the original recommendations. Recommendations that have been deleted or changed sets out these recommendations and includes details of replacement recommendations. Where there is no replacement recommendation, an explanation for the proposed deletion is given.

Where recommendations are shaded in grey and end **[2010]**, the evidence has not been reviewed since the original guideline.

Where recommendations are shaded in grey and end **[2010, amended 2016]**, the evidence has not been reviewed but changes have been made to the recommendation wording that change the meaning (for example, because of equalities duties or a change in the availability of medicines, or incorporated guidance has been updated). These changes are marked with yellow shading,

and explanations of the reasons for the changes are given in 'Recommendations that have been deleted or changed' for information.

See also the original NICE guideline and supporting documents.

Recommendations that have been deleted or changed

Recommendations to be deleted

The table shows recommendations from 2010 that NICE proposes deleting in the 2016 update. The right-hand column gives the replacement recommendation, or explains the reason for the deletion if there is no replacement recommendation.

Recommendation in 2010 guideline	Comment
A multidisciplinary team should coordinate and provide ongoing management and treatment for a patient with MND, including regular respiratory assessment and provision of non-invasive ventilation. • The team should be led by a healthcare professional with a specific interest in MND. The leader should ensure that the patient's multidisciplinary care plan (see recommendation 1.1.19) is coordinated and is communicated to relevant healthcare and social care professionals, including the patient's primary care team, as well as to the patient and (where appropriate) their family and carers. • The team should include a neurologist, a respiratory physician, an MND specialist nurse, a respiratory specialist nurse, a specialist respiratory physiotherapist, a respiratory physiologist, a specialist in palliative care and a speech and language therapist (team members do not have to be at the same location). • Access to other healthcare professionals should be provided as needed. • Team members who provide non-invasive ventilation should have appropriate competencies. (1.1.1)	Replaced by: 1.5.1 Provide coordinated care for people with MND, using a clinic-based, multidisciplinary team approach. [new 2016] 1.5.2 The multidisciplinary team should: • include healthcare professionals and social care practitioners with expertise in MND, and staff who see people in their home • ensure effective communication between all healthcare professionals and social care practitioners involved in the person's care and their family members and/or carers (as appropriate) • carry out regular, coordinated assessments at the multidisciplinary team clinic (usually every 2–3 months) to assess people's symptoms and needs. [new 2016] 1.5.3 The multidisciplinary team should assess the following: • Weight, diet, nutritional intake, feeding and swallowing (see recommendations 1.10.1–1.10.10). • Muscle problems, such as weakness, stiffness, cramps (see recommendations 1.8.1–1.8.9).

	• Physical function, including mobility and activities of daily living (see recommendations 1.9.1–1.9.8). • Saliva problems, such as drooling of saliva (sialorrhoea) and thick, tenacious saliva (see recommendations 1.8.10–1.8.15). • Speech and communication (see recommendations 1.11.1–1.11.6). • Cough effectiveness (see recommendations 1.12.1–1.12.4). • Respiratory function (see section 1.13). • Pain and other symptoms, such as constipation. • Cognition and behaviour (see recommendations 1.3.1–1.3.3). • Psychological support needs (see recommendations 1.6.1–1.6.4). • Social care needs (see recommendations 1.6.5–1.6.6). • Information and support needs for the person and their family members and/or carers (as appropriate). [new 2016] 1.5.4 The core multidisciplinary team should consist of healthcare professionals and other professionals with expertise in MND, and should include the following: • Neurologist. • Specialist nurse. • Dietitian. • Physiotherapist. • Occupational therapist. • Respiratory physiologist or a healthcare professional who can assess respiratory function. • Speech and language therapist. [new 2016] 1.5.5 The multidisciplinary team should have access to the following services: • Clinical psychology and/or neuropsychology. • Social care. • Counselling. • Respiratory medicine. • Specialist palliative care. • Gastroenterology. [new 2016]

	1.5.6 Tailor the frequency of the multidisciplinary team assessments to the person's symptoms and needs, with more or less frequent assessments as needed. [new 2016] 1.5.7 Ensure arrangements are in place to trigger an earlier multidisciplinary team assessment if there is a significant change in symptoms identified by family members and/or carers (as appropriate), or healthcare professionals. [new 2016] 1.5.8 Tailor the multidisciplinary team assessment to the person's needs, for example, adjust the format if the person has cognitive or behaviour changes or difficulties with communication. [new 2016] 1.5.9 Inform all healthcare professionals and social care practitioners involved in the person's care about key decisions reached with the person and their family members and/or carers (as appropriate). [new 2016] 1.5.10 Ensure that all healthcare professionals and social care practitioners involved in the person's care are aware that MND symptoms may get worse quickly, and that people with MND will need repeated assessment. Priority should be given to ensuring continuity of care and avoidance of untimely case closure. [new 2016]
Inform all relevant healthcare professionals about key decisions reached with the patient and their family and carers. (1.1.4)	This recommendation has been deleted because similar recommendation in new guideline. 1.5.9 Inform all healthcare professionals and social care practitioners involved in the person's care about key decisions reached with the person and their family members and/or carers (as appropriate). [new 2016]
1.1.24 Offer to discuss end-of-life care with the patient and (if the patient agrees) their family and carers, at an appropriate time and in a sensitive manner. This may be at one or more of	Deleted and replaced with recommendations on Planning for end of life. 1.7.1 Offer the person with MND the opportunity to discuss their preferences

the following times: • around the time that MND is first diagnosed (but only if requested by the patient explicitly, or if the patient's clinical condition indicates that ventilator support will be needed in the immediate future) • when non-invasive ventilation is accepted or declined • when the patient is becoming increasingly dependent on non-invasive ventilation • if the patient asks for information.	and concerns about care at the end of life at trigger points such as: at diagnosis, if there is a significant change in respiratory function, or if interventions such as gastrostomy or non-invasive ventilation are needed. Be sensitive about the timing of discussions and take into account the person's current communication ability, cognitive status and mental capacity. [new 2016] 1.7.2 Think about discussing advance care planning with people at an earlier opportunity if you expect their communication ability, cognitive status or mental capacity to get worse. [new 2016] 1.7.3 Consider an early referral to a specialist palliative care team for people with significant or complex needs, such as psychological or social distress or rapidly progressing symptoms. [new 2016] 1.7.4 Provide support and advice on advance care planning for end of life to the person with MND and their family members and/or carers (as appropriate). The discussion should include: • What could happen at end of life, for example how death may occur. • Providing anticipatory medicines in the home. • Advance care planning, including Advanced Decisions to Refuse Treatment (ADRT) and Do Not Attempt Resuscitation (DNACPR) orders, and Lasting Power of Attorney. • Areas that people might wish to plan for, such as: – what they want to happen (for example preferred place of death) – what they do not want to happen (for example being admitted to hospital) – who will represent their decisions, if necessary – what should happen if they develop an intercurrent illness. [new 2016] 1.7.5 Offer people the opportunity to talk about ADRT, DNACPR and Lasting Power of Attorney when interventions

	such as gastrostomy and non-invasive ventilation are planned. [new 2016] 1.7.6 Provide additional support as the end of life approaches, for example, additional social or nursing care to enable informal carers and family to reduce their carer responsibilities and spend time with the person with MND. [new 2016] 1.7.7 Towards the end of life, ensure there is access to the following: • An appropriate method of communication, such as an alternative and augmentative communication (AAC) system. • Holistic support. • Specialist palliative care. • Equipment, if needed, such as syringe drivers, suction machines, riser–recliner chair, hospital bed, commode, hoist. • Anticipatory medicines including opioids and benzodiazepines to treat breathlessness, and anticholinergic medicines to treat problematic saliva and respiratory secretions. [new 2016] 1.7.8 Offer bereavement support to family members and/or carers (as appropriate). [new 2016]
1.1.25 Discussions about end-of-life care should include: • planning of end-of-life care • considering advance decisions to refuse treatment • considering what to do if non-invasive ventilation fails because of either: – an acute, but potentially reversible, deterioration in health or – irreversible disease progression • strategies to withdraw non-invasive ventilation if the patient wishes • the involvement of family and carers in decision making (with the patient's consent if they have the capacity to give it).	Deleted and replaced with recommendations on: 1.Planning for end of life (section 1.7) 2. Information and support about non-invasive ventilation (recommendations 1.14.2–1.14.5) 3. Stopping non-invasive ventilation (recommendations 1.14.28–1.14.31)

1

Amended recommendation wording (change to meaning)

Recommendations are labelled **[2010, amended 2016]** if the evidence has not been reviewed but changes have been made to the recommendation wording (indicated by highlighted text) that change the meaning.

Recommendation in 2010 guideline	Recommendation in current guideline	Reason for change
Discussions should be appropriate to the stage of the patient's illness, carried out in a sensitive manner and include information on: • the possible symptoms and signs of respiratory impairment (see table 1 in recommendation 1.1.7) • the natural progression of MND and what to expect in the future • the purpose, nature and timing of respiratory function tests, and explanations of the test results • available interventions for managing respiratory impairment, including the benefits and limitations of each intervention • accessing and using respiratory equipment, including that for non-invasive ventilation • how non-invasive ventilation (as a treatment option) can improve symptoms associated with respiratory impairment and can be life prolonging, but does not stop progression of the underlying disease • how non-invasive ventilation can be withdrawn • palliative strategies as an alternative to non-invasive ventilation. (1.1.3)	1.14.2 Discussions about non-invasive ventilation should be appropriate to the stage of the person's illness, carried out in a sensitive manner and include information on: • the possible symptoms and signs of respiratory impairment (see table 1) • the purpose, nature and timing of respiratory function tests, and explanations of the test results • how non-invasive ventilation (as a treatment option) can improve symptoms associated with respiratory impairment and can be life prolonging, but does not stop progression of the underlying disease. [2010, amended 2016]	Amended to include specific reference to non-invasive ventilation, and to ensure consistency with new recommendations on information about non-invasive ventilation.
1.1.5 Provide the patient and their family and carers	1.14.23 Provide the person and their family and/or carers	Footnote removed as psychological and

with support and assistance to manage non-invasive ventilation. This should include: • training on using non-invasive ventilation and ventilator interfaces, for example: – emergency procedures – night-time assistance if the patient is unable to use the equipment independently (for example, emergency removal or replacement of interfaces) – how to use the equipment with a wheelchair or other mobility aids if required – what to do if the equipment fails • assistance with secretion management • information on general palliative strategies • an offer of ongoing emotional and psychological support[1] for the patient and their family and carers.	(as appropriate) with support and assistance to manage non-invasive ventilation. This should include: • training on using non-invasive ventilation and ventilator interfaces, for example: – emergency procedures – night-time assistance if the person is unable to use the equipment independently (for example, emergency removal or replacement of interfaces) – how to use the equipment with a wheelchair or other mobility aids if required – what to do if the equipment fails • assistance with secretion management • information on general palliative strategies • an offer of ongoing emotional and psychological support for the person and their family and carers. [2010, amended 2016]	social support are included in the new guideline.
1.1.7 Monitor the symptoms and signs listed in table 1 routinely to detect potential respiratory impairment.	1.14.7 Monitor the symptoms and signs listed in table 1 to detect potential respiratory impairment. [2010, amended 2016]	'Routinely' removed as new recommendations for the multidisciplinary team advise on frequency of assessment.
1.1.10 A healthcare professional with appropriate competencies should perform the respiratory function tests every 3 months, although tests may be performed more or less often depending on: • whether there are any symptoms and signs of respiratory impairment	1.14.10 A healthcare professional with appropriate competencies should perform the respiratory function tests every 2–3 months, although tests may be performed more or less often depending on: • whether there are any symptoms and signs of respiratory impairment	Time period removed as already included in recommendations for the multidisciplinary team.

(see recommendation 1.1.7) • the rate of progression of MND • the patient's preference and circumstances.	(see table 1) • the rate of progression of MND • the person's preference and circumstances. [2010, amended 2016]	
1.1.15 If any of the results listed in table 2 is obtained, discuss with the patient and (if the patient agrees) their family and carers: • the impact of respiratory impairment • treatment options • possible referral to a specialist respiratory service for further assessment.	1.14.15 If any of the results listed in table 2 is obtained, discuss with the person and (if appropriate) their family and carers: • their respiratory impairment • their treatment options • possible referral to a specialist respiratory service for further assessment based on discussion with the person, and their wishes. [2010, amended 2016]	Wording changed for consistency and to emphasise patient choice for referral.
1.1.16 Base decisions on respiratory function tests for a patient with a diagnosis of dementia on considerations specific to their needs and circumstances, such as: • their ability to give consent[3] • their understanding of the tests • their tolerance of the tests and willingness to undertake them • the impact on their family and carers • whether they are capable of receiving non-invasive ventilation.	1.14.16 Base decisions on respiratory function tests for a person with a diagnosis of frontotemporal dementia on considerations specific to their needs and circumstances, such as: • their ability to give consent[6] • their understanding of the tests • their tolerance of the tests and willingness to undertake them • the impact on their family and carers • whether they are capable of receiving non-invasive ventilation. [2010, amended 2016] [6] See Mental Capacity Act 2005.	Amended for consistency without change in meaning. 'dementia' changed to 'frontotemporal dementia'. Footnote wording has changed to refer to the Mental Capacity Act 2005
1.1.17 Offer a trial of non-invasive ventilation if the patient's symptoms and signs and the results of the respiratory function tests indicate that the patient is likely to benefit from the treatment.	1.14.17 Offer a trial of non-invasive ventilation if the person's symptoms and signs and the results of the respiratory function tests indicate that the person is likely to benefit from the treatment. [2010, amended	Amended for consistency with new recommendations. New recommendations developed for information following evidence review on

• Discuss both the benefits and limitations of the intervention with the patient and their family and carers. • Only consider a trial of non-invasive ventilation for a patient who has severe bulbar impairment or severe cognitive problems that may be related to respiratory impairment if they may benefit from an improvement in sleep-related symptoms or correction of hypoventilation.	2016] 1.14.18 Consider a trial of non-invasive ventilation for a person who has severe bulbar impairment or severe cognitive problems that may be related to respiratory impairment only if they may benefit from an improvement in sleep-related symptoms or correction of hypoventilation. [2010, amended 2016]	stopping non-invasive ventilation.
1.1.19 Before starting non-invasive ventilation, the multidisciplinary team should prepare a comprehensive care plan, after discussion with the patient and their family and carers (who should be offered a copy of the plan). This should cover: • long-term support provided by the multidisciplinary team • the initial frequency of respiratory function tests and monitoring of respiratory impairment • the frequency of clinical reviews of symptomatic and physiological changes • the provision of carers • arrangements for device maintenance and 24-hour emergency clinical and technical support • secretion management and respiratory physiotherapy assessment, including cough-assist therapy (if required) • training in and support for the use of non-invasive ventilation for the patient and their family and carers	1.14.20 Before starting non-invasive ventilation, the multidisciplinary team should prepare a comprehensive care plan, after discussion with the person and their family and carers (who should be offered a copy of the plan). This should cover: • long-term support provided by the multidisciplinary team • the initial frequency of respiratory function tests and monitoring of respiratory impairment • the frequency of clinical reviews of symptomatic and physiological changes • the provision of carers • arrangements for device maintenance and 24-hour emergency clinical and technical support • secretion management and respiratory physiotherapy assessment, including cough-assist therapy (if required) • training in and support for the use of non-invasive ventilation for the person and their family and carers	Part of last bullet point deleted as replaced by recommendations on Planning for end of life.

• regular opportunities to discuss the patient's wishes in relation to continuing or withdrawing non-invasive ventilation, and other end-of-life considerations (see also recommendations 1.1.24 and 1.1.25).	• regular opportunities to discuss the person's wishes in relation to continuing or withdrawing non-invasive ventilation. [2010, amended 2016]	
1.1.23 Before a decision is made on the use of non-invasive ventilation for a patient with a diagnosis of dementia, the neurologist from the multidisciplinary team should carry out an assessment that includes: • the patient's capacity to make decisions and to give consent[3] • the severity of dementia and cognitive problems • whether the patient is likely to accept treatment • whether the patient is likely to achieve improvements in sleep-related symptoms and/or behavioural improvements • a discussion with the patient's family and/or carers (with the patient's consent if they have the capacity to give it).	1.14.25 Before a decision is made on the use of non-invasive ventilation for a person with a diagnosis of frontotemporal dementia, the multidisciplinary team should carry out an assessment that includes: • the person's capacity to make decisions and to give consent[7] • the severity of dementia and cognitive problems • whether the person is likely to accept treatment • whether the person is likely to achieve improvements in sleep-related symptoms and/or behavioural improvements • a discussion with the person's family and/or carers (with the person's consent if they have the capacity to give it). [2010, amended 2016] [7]See Mental Capacity Act 2005.	Amended to update wording and reflect changes to law. 'dementia' changed to 'frontotemporal dementia' and footnote changed to reflect Mental Capacity Act. Footnote wording has changed to refer to the Mental Capacity Act 2005.

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