

## Motor neurone disease overview

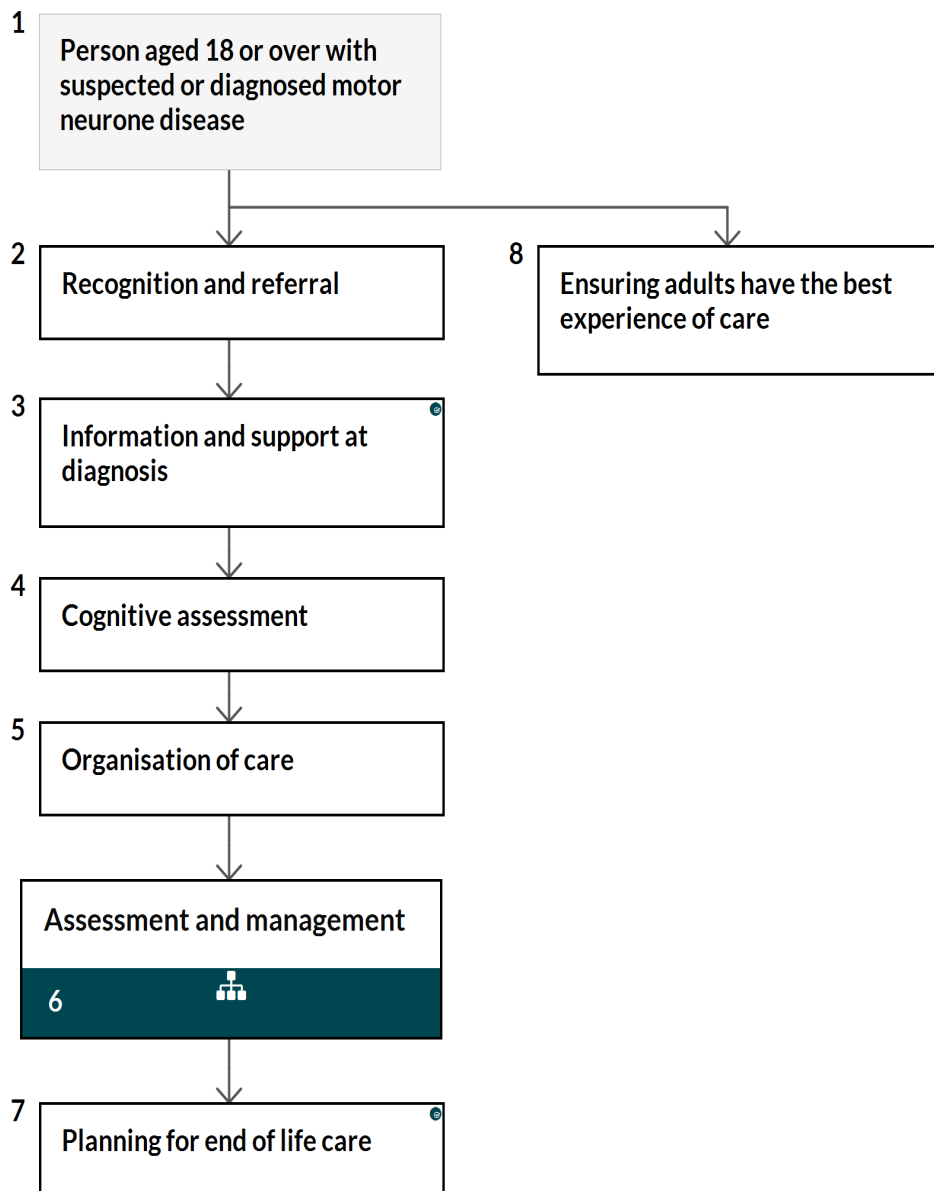
NICE Pathways bring together everything NICE says on a topic in an interactive flowchart. NICE Pathways are interactive and designed to be used online.

They are updated regularly as new NICE guidance is published. To view the latest version of this NICE Pathway see:

<http://pathways.nice.org.uk/pathways/motor-neurone-disease>

NICE Pathway last updated: 07 August 2020

This document contains a single flowchart and uses numbering to link the boxes to the associated recommendations.



## 1 Person aged 18 or over with suspected or diagnosed motor neurone disease

No additional information

## 2 Recognition and referral

Ensure that robust protocols and pathways are in place to:

- inform healthcare professionals about motor neurone disease and how it may present
- inform healthcare professionals in all settings about local referral arrangements
- ensure continued and integrated care for people with motor neurone disease across all care settings.

Be aware that motor neurone disease 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.

Be aware that motor neurone disease may first present with cognitive features, which may include:

- behavioural changes
- emotional lability (not related to dementia)
- frontotemporal dementia.

If you suspect motor neurone disease, refer the person without delay and specify the possible diagnosis in the referral letter. Contact the consultant neurologist directly if you think the person needs to be seen urgently.

Provide information and support for people and their family members and/or carers (as appropriate) throughout the diagnostic process, particularly during periods of diagnostic

uncertainty or delay.

### 3 Information and support at diagnosis

Please also refer to [the NICE Pathway on patient experience in adult NHS services](#), which includes recommendations on communication, information and coordination of care.

Information about the diagnosis, prognosis and management of motor neurone disease should be given by a consultant neurologist with up-to-date knowledge and experience of treating people with motor neurone disease 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 motor neurone disease.
- Types and possible causes of motor neurone disease.
- Treatment options.
- How motor neurone disease 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 motor neurone disease to be involved in research.
- Likely needs and concerns of people with motor neurone disease and their family members and/or carers (as appropriate).
- Advance care planning.

Ask people about how much information they wish to receive about motor neurone disease, and about their preferences for involving their family members and/or carers (as appropriate).

Ensure people are provided with information and support about motor neurone disease at diagnosis or when they 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 may include the following:

- What motor neurone disease is.
- Types and possible causes.
- Likely symptoms and how they can be managed.
- How motor neurone disease 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 and specialist palliative 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.
- Opportunities for advance care planning.

When motor neurone disease is diagnosed, provide people with a single point of contact for the specialist motor neurone disease multidisciplinary team (see [organisation of care \[See page 6\]](#)). Provide 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.

Offer the person with motor neurone disease a face-to-face, follow-up appointment with a healthcare professional from the multidisciplinary team, to take place within 4 weeks of diagnosis.

When motor neurone disease is suspected or confirmed, inform the person's GP without delay and provide information about the likely prognosis.

Set aside enough time to discuss the person's concerns and questions, 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 motor neurone disease?
- How do I tell my family and friends?
- How will I die?

If the person has any social care needs, refer them to social services for an assessment. Be aware that some people with motor neurone disease may not have informal care available, and

may live alone or care for someone else.

Advise carers about their right to carer assessment, and assessment for respite care and other support (see [the NICE Pathway on supporting adult carers](#) for recommendations on identifying, assessing and meeting the caring, physical and mental health needs of families and carers).

NICE has written [information for the public on motor neurone disease](#).

## Quality standards

The following quality statement is relevant to this part of the interactive flowchart.

### 1. Information and support at diagnosis

#### 4 Cognitive assessment

Please also refer to [the NICE Pathway on patient experience in adult NHS services](#).

Be aware that people with motor neurone disease and frontotemporal dementia may lack mental capacity. Care should be provided in line with the [Mental Capacity Act 2005](#).

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 Pathway on dementia assessment and diagnosis](#).

Tailor all discussions to the person's needs, taking into account their communication ability, cognitive status and mental capacity.

#### 5 Organisation of care

When planning care 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.

Provide coordinated care for people with motor neurone disease, using a clinic-based, specialist motor neurone disease multidisciplinary team approach. The clinic may be community or hospital based.

The multidisciplinary team should:

- include healthcare professionals and social care practitioners with expertise in motor neurone disease, and staff who see people in their home
- ensure effective communication and coordination 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.
- provide coordinated care for people who cannot attend the clinic, according to the person's needs.

The core multidisciplinary team should consist of healthcare professionals and other professionals with expertise in motor neurone disease, 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.
- A healthcare professional with expertise in palliative care (motor neurone disease palliative care expertise may be provided by the neurologist or nurse in the multidisciplinary team, or by a specialist palliative care professional).

The multidisciplinary team should have established relationships with, and prompt access to, the following:

- Clinical psychology and neuropsychology.
- Social care.
- Counselling.

- Respiratory ventilation services.
- Specialist palliative care.
- Gastroenterology.
- Orthotics.
- Wheelchair services.
- Assistive technology services.
- AAC services.
- Community neurological care teams.

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).

Ensure that all healthcare professionals and social care practitioners involved in the person's care are aware that motor neurone disease symptoms may get worse quickly, and that people with motor neurone disease will need repeated, ongoing assessments. Priority should be given to ensuring continuity of care and avoiding untimely case closure.

Consider referral to a specialist palliative care team for people with current or anticipated significant or complex needs, for example, psychological or social distress, troublesome or rapidly progressing symptoms and complex future care planning needs.

## 6 Assessment and management

[See Motor neurone disease / Assessing and managing motor neurone disease](#)

## 7 Planning for end of life care

Offer the person with motor neurone disease 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.

Be prepared to discuss end of life issues whenever people wish to do so.

Provide support and advice on advance care planning for end of life. Topics to discuss may



include:

- What could happen at the end of life, for example, how death may occur.
- Providing anticipatory medicines in the home.
- Advance care planning, including ADRT and DNACPR orders, and Lasting Power of Attorney.
- How to ensure advance care plans will be available when needed, for example, including the information on the person's Summary Care Record.
- When to involve specialist palliative care.
- Areas that people might wish to plan for, such as:
  - what they want to happen (for example, their 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.

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.

Offer people the opportunity to talk about, and review any existing ADRT or DNACPR orders and Lasting Power of Attorney when interventions such as gastrostomy and non-invasive ventilation are planned.

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 motor neurone disease.

Towards the end of life, ensure there is prompt access to the following, if not already provided:

- A method of communication that meets the person's needs, such as an AAC system.
- Specialist palliative care.
- Equipment, if needed, such as syringe drivers, suction machines, riser–recliner chair, hospital bed, commode and hoist.
- Anticipatory medicines, including opioids and benzodiazepines to treat breathlessness, and antimuscarinic medicines to treat problematic saliva and respiratory secretions.

Offer bereavement support to family members and/or carers (as appropriate).

See [NICE's recommendations on caring for an adult at the end of life](#).

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## Quality standards

The following quality statement is relevant to this part of the interactive flowchart.

5. Planning for end of life care

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### Experience of care

Use these recommendations together with the recommendations in the NICE Pathways on:

- patient experience in adult NHS services
- people's experience in adult social care services.

## Glossary

### AAC

augmentative and alternative communication

### ADRT

advance decisions to refuse treatment

### DNACPR

do not attempt resuscitation

## Sources

[Motor neurone disease: assessment and management \(2016\) NICE guideline NG42](#)

## Your responsibility

### Guidelines

The recommendations in this guideline represent the view of NICE, arrived at after careful consideration of the evidence available. When exercising their judgement, professionals and practitioners are expected to take this guideline fully into account, alongside the individual needs, preferences and values of their patients or the people using their service. It is not mandatory to apply the recommendations, and the guideline does not override the responsibility to make decisions appropriate to the circumstances of the individual, in consultation with them and their families and carers or guardian.

Local commissioners and providers of healthcare have a responsibility to enable the guideline to be applied when individual professionals and people using services wish to use it. They should do so in the context of local and national priorities for funding and developing services, and in light of their duties to have due regard to the need to eliminate unlawful discrimination, to advance equality of opportunity and to reduce health inequalities. Nothing in this guideline should be interpreted in a way that would be inconsistent with complying with those duties.

Commissioners and providers have a responsibility to promote an environmentally sustainable health and care system and should assess and reduce the environmental impact of implementing NICE recommendations wherever possible.

### **Technology appraisals**

The recommendations in this interactive flowchart represent the view of NICE, arrived at after careful consideration of the evidence available. When exercising their judgement, health professionals are expected to take these recommendations fully into account, alongside the individual needs, preferences and values of their patients. The application of the recommendations in this interactive flowchart is at the discretion of health professionals and their individual patients and do not override the responsibility of healthcare professionals to make decisions appropriate to the circumstances of the individual patient, in consultation with the patient and/or their carer or guardian.

Commissioners and/or providers have a responsibility to provide the funding required to enable the recommendations to be applied when individual health professionals and their patients wish to use it, in accordance with the NHS Constitution. They should do so in light of their duties to have due regard to the need to eliminate unlawful discrimination, to advance equality of opportunity and to reduce health inequalities.

Commissioners and providers have a responsibility to promote an environmentally sustainable health and care system and should assess and reduce the environmental impact of implementing NICE recommendations wherever possible.

### **Medical technologies guidance, diagnostics guidance and interventional procedures guidance**

The recommendations in this interactive flowchart represent the view of NICE, arrived at after careful consideration of the evidence available. When exercising their judgement, healthcare professionals are expected to take these recommendations fully into account. However, the interactive flowchart does not override the individual responsibility of healthcare professionals to make decisions appropriate to the circumstances of the individual patient, in consultation with the patient and/or guardian or carer.

Commissioners and/or providers have a responsibility to implement the recommendations, in

their local context, in light of their duties to have due regard to the need to eliminate unlawful discrimination, advance equality of opportunity, and foster good relations. Nothing in this interactive flowchart should be interpreted in a way that would be inconsistent with compliance with those duties.

Commissioners and providers have a responsibility to promote an environmentally sustainable health and care system and should assess and reduce the environmental impact of implementing NICE recommendations wherever possible.