NATIONAL INSTITUTE FOR HEALTH AND CARE EXCELLENCE

Health and social care directorate

Quality standards and indicators

Briefing paper

Quality standard topic: Motor neurone disease

Output: Prioritised quality improvement areas for development.

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Contents

1 Introduction

This briefing paper presents a structured overview of potential quality improvement areas for motor neurone disease. It provides the Committee with a basis for discussing and prioritising quality improvement areas for development into draft quality statements and measures for public consultation.

1.1 Structure

This briefing paper includes a brief description of the topic, a summary of each of the suggested quality improvement areas and supporting information.

If relevant, recommendations selected from the key development source below are included to help the Committee in considering potential statements and measures.

1.2 Development source

The key development source referenced in this briefing paper is:

- Motor Neurone Disease. NICE clinical guideline in development (draft for consultation). Anticipated publication date February 2016

2 Overview

2.1 Focus of quality standard

This quality standard will cover the assessment and management of motor neurone disease.

2.2 Definition

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.

2.3 Incidence and prevalence

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.

2.4 Management

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. MND Care Centres can provide multidisciplinary care.

2.5 National Outcome Frameworks

Tables 1 and 2 show the outcomes, overarching indicators and improvement areas from the frameworks that the quality standard could contribute to achieving.

Table 1 The Adult Social Care Outcomes Framework 2015–16

Domain	Overarching and outcome measures
1 Enhancing quality of life for people with care and support needs	***Overarching measure*** 1A Social care-related quality of life** ***Outcome measures*** **People manage their own support as much as they wish, so they are in control of what, how and when support is delivered to match their needs** 1B Proportion of people who use services who have control over their daily life **Carers can balance their caring roles and maintain their desired quality of life** 1D Carer-reported quality of life**
3 Ensuring that people have a positive experience of care and support	***Overarching measure*** **People who use social care and their carers are satisfied with their experience of care and support services** 3A Overall satisfaction of people who use services with their care and support 3B Overall satisfaction of carers with social services *Placeholder 3E The effectiveness of integrated care* ***Outcome measures*** **Carers feel that they are respected as equal partners throughout the care process** 3C The proportion of carers who report that they have been included or consulted in discussions about the person they care for **People know what choices are available to them locally, what they are entitled to, and who to contact when they need help** 3D The proportion of people who use services and carers who find it easy to find information about support **People, including those involved in making decisions on social care, respect the dignity of the individual and ensure support is sensitive to the circumstances of each individual** This information can be taken from the Adult Social Care Survey and used for analysis at the local level.

4 Safeguarding adults whose circumstances make them vulnerable and protecting from avoidable harm	***Overarching measure*** 4A The proportion of people who use services who feel safe** ***Outcome measures*** **Everyone enjoys physical safety and feels secure** **People are free from physical and emotional abuse, harassment, neglect and self-harm** **People are protected as far as possible from avoidable harm, disease and injuries** **People are supported to plan ahead and have the freedom to manage risks the way that they wish** 4B The proportion of people who use services who say that those services have made them feel safe and secure *Placeholder 4C Proportion of completed safeguarding referrals where people report they feel safe*
Alignment with NHS Outcomes Framework and/or Public Health Outcomes Framework * Indicator is shared ** Indicator is complementary Indicators in italics in development	

Table 2 NHS Outcomes Framework 2015–16

Domain	Overarching indicators and improvement areas
2 Enhancing quality of life for people with long-term conditions	***Overarching indicator*** 2 Health-related quality of life for people with long-term conditions** ***Improvement areas*** **Ensuring people feel supported to manage their condition** 2.1 Proportion of people feeling supported to manage their condition **Reducing time spent in hospital by people with long-term conditions** 2.3 i Unplanned hospitalisation for chronic ambulatory care sensitive conditions **Enhancing quality of life for carers** 2.4 Health-related quality of life for carers** **Enhancing quality of life for people with mental illness** *ii Health-related quality of life for people with mental illness***
4 Ensuring that people have a positive experience of care	***Overarching indicators*** 4b Patient experience of hospital care 4c *Friends and family test* *4d Patient experience characterised as poor or worse* *I Primary care* *ii Hospital care* ***Improvement areas*** **Improving the experience of care for people at the end of their lives** 4.6 Bereaved carers' views on the quality of care in the last 3 months of life **Improving people's experience of integrated care** *4.9 People's experience of integrated care***
Alignment with Adult Social Care Outcomes Framework and/or Public Health Outcomes Framework * Indicator is shared ** Indicator is complementary Indicators in italics in development	

3 Summary of suggestions

3.1 Responses

In total 9 stakeholders responded to the engagement exercise [01/09/15 to 13/10/15].

Stakeholders were asked to suggest up to 5 areas for quality improvement. Specialist committee members were also invited to provide suggestions. The responses have been merged and summarised in table 3 for further consideration by the Committee.

Full details of all the suggestions provided are given in appendix 2 for information.

Table 3 Summary of suggested quality improvement areas

Suggested area for improvement	Stakeholders
Diagnosis • Recognition and referral • Information and support at diagnosis • Cognitive assessments	BSRM, MNDA, RCP, SCM4, SCM 5
Organisation of care • Multidisciplinary teams • Social care support	BSRM, MNDA, RDS NHS, SCM2, SCM3, SCM4, SCM5
Respiratory impairment and cough effectiveness • Assessment of respiratory function • Cough effectiveness • Provision of non-invasive ventilation (NIV)	BSRM, SCM1, SCM2, SCM3, SCM5
Planning for end of life	SCM4, SCM5
Provision of equipment • Equipment and adaptations to aid activities of daily living and mobility • Augmentative and Alternative Communication (AAC) • Wheelchairs • Distress/crisis interventions	MNDA, RDS NHS, SCM2, SCM3, SCM4, SCM5
Additional areas • Riluzole • Genetic counselling	MNDA, RCP, SCM3
BSRM, British Society of Rehabilitation Medicine MNDA, Motor Neurone Disease Association RCP, Royal College of Pathologists RDS NHS, Rotherham Doncaster and South Humber NHS FT SCM, Specialist Committee Member	

3.2 Identification of current practice evidence

Bibliographic databases were searched to identify examples of current practice in UK health and social care settings; 429 studies were identified for QS topic. In addition, current practice examples were suggested by stakeholders at topic engagement (5 studies) and internally at project scoping (12 studies).

Of these studies, 10 were assessed as having potential relevance to this topic and the suggested areas for quality improvement identified by stakeholders (see appendix 1). A summary of relevant studies is included in the current practice sections for each suggested area of improvement.

4 Suggested improvement areas

4.1 Diagnosis

4.1.1 Summary of suggestions

Recognition and referral

Several stakeholders highlighted difficulties and delays in diagnosing motor neurone disease (MND) - a rare disease for which there is no diagnostic test. A stakeholder commented that securing a referral from a GP to a neurologist is the first major hurdle to getting a diagnosis, commenting that people are often inappropriately referred to other specialists. A stakeholder also commented that there is a need for diagnostic pathways to be put in place to expedite diagnosis.

Information and support at diagnosis

Stakeholders also emphasised how distressing a diagnosis of MND can be and highlighted the importance of breaking news in a considerate manner. For example, being advised to bring a friend or relative, being given a diagnosis in a private room and being given the opportunity to compose themselves after receiving a diagnosis.

A stakeholder commented that it is important that people, and their families, get the right information and support at diagnosis to ensure that they are informed and able to access care. The stakeholder also commented that most GPs and healthcare professionals lack the appropriate knowledge of MND to carry this out; a specialist neurologist with appropriate MND experience is required to lead this process.

A stakeholder also suggested that people with MND should have the opportunity to participate in research, commenting that improvements in treatment and identifying new biomarkers for diagnosis will only occur with the participation of patients.

Cognitive assessments

Stakeholders highlighted the importance of carrying out cognitive assessment for people with MND, because this will impact on how their symptoms are managed and how interventions/equipment are used.

4.1.2 Selected recommendations from development source

Table 4 below highlights recommendations that have been provisionally selected from the development source(s) that may support potential statement development. These are presented in full after table 4 to help inform the Committee’s discussion.

Table 4 Specific areas for quality improvement

Suggested quality improvement area	Suggested source guidance recommendations
Recognition and referral	**Recognition and referral** Motor Neurone Disease. NICE clinical guideline in development. Recommendation 1.1.4
Information and support at diagnosis	**Information and support at diagnosis** Motor Neurone Disease. NICE clinical guideline in development. Recommendation 1.2.1 Recommendation 1.2.5
Cognitive assessments	**Cognitive assessment** Motor Neurone Disease. NICE clinical guideline in development. Recommendation 1.3.2

Recognition and referral

Motor Neurone Disease. NICE clinical guideline in development – Recommendation 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.

Information and support at diagnosis

Motor Neurone Disease. NICE clinical guideline in development – Recommendation 1.2.1

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.

Motor Neurone Disease. NICE clinical guideline in development – Recommendation 1.2.5

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.

Cognitive assessment

Motor Neurone Disease. NICE clinical guideline in development - Recommendation 1.3.2

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.

4.1.3 Current UK practice

Recognition and referral

A study published in 2008 commented that GPs would expect to see one or two cases of motor neurone disease during their career[1].

[1] Diagnosis and management of motor neurone disease (2008) BMJ. 2008 336(7645): 658–662.

A study of patients diagnosed with MND at the Preston MND Care Facility between 1 January 1989 and 31 December 2008 (n=640) reported that the median time from first symptom to diagnosis was 0.95 years (95% CI 0.21-5.01)[2]. The authors reported that this time showed consistency across the 20 year time span of the study. The authors also identified three reasons for delayed diagnosis: (1) patients presenting with atypical symptoms (which are inconclusive or resemble disorders other than MND), (2) delays in primary care recognition of MND symptoms, and (3) patients were being referred to specialist hospital departments other than neurology.

The 2013 MND Association 'Improving MND Care' survey[3] collated responses from 951 people with MND[4] and reported on several topics relevant to diagnosis, including:

- From their first consultation with a GP to being referred to a neurologist:
    - 60% were referred within 6 months
    - 17% waited for a year or more

When asked about their referral to a neurologist, 32% respondents replied that this did not occur as soon as they would have liked (37% replied that referral was definitely as soon as they would have liked and 27% replied that it had 'to some extent').

The survey also reported that 45% of respondents were referred directly to a neurologist and (after having been referred) 76% of respondents were seen within 3 months.

A cross sectional survey involving patient members of the Motor Neurone Disease Association (505 respondents included in study) investigated patient experience of health and social care related to MND in England between October 2008 and January 2009[5]. The study reported that:

- the time from GP consultation to specialist consultation was more than 6 months for almost 36% of respondents
- The time from first GP consultation to diagnosis was more than 12 months for 44% respondents

[2] Timelines in the diagnostic evaluation of people with suspected amyotrophic lateral sclerosis (ALS)/motor neuron disease (MND) – a 20-year review: Can we do better? (2010) Amyotrophic Lateral Sclerosis 11, 537-541
[3] Improving MND Care (2013) MND Association
[4] Respondents to the survey had received a confirmed diagnosis of MND within the last year (30%), between one and three years previously (33%) or more than three years previously (37%).
[5] Patients' experiences of health and social care in long-term neurological conditions in England: a cross-sectional survey (2013) Peters M, Fitzpatrick R, Doll H, Playford ED and Jenkinson C. J Health Serv Res Policy January 2013 vol. 18 no. 1 28-33

Information and support at diagnosis

The MND Association's 'Improving MND Care' survey reported that 93% of respondents completing the survey received their diagnosis '*in person, during an appointment with a neurologist*'[6]. In addition, 5% respondents received their diagnoses during an appointment with another doctor or healthcare professional, 2% received their diagnosis by letter and less than 0.5% received theirs by telephone.

In addition, also from this survey:

- 79% of respondents felt that they had been given the opportunity to ask questions and a further 16% felt that they had been given this opportunity 'to some extent'. 3% of respondents would have liked to have been given the opportunity to ask questions but were not offered this opportunity.

- 88% of respondents felt that the person giving the diagnosis definitely had a good understanding of MND, and a further 9% agreed that this was the case 'to some extent'.

- 59% of respondents 'definitely' felt supported throughout their consultation. 17% of respondents said that they did not feel supported (and either did or did not mind).

A cross sectional survey involving patient members of the Motor Neurone Disease Association (505 respondents included in study) investigated patient experience of health and social care related to MND in England between October 2008 and January 2009[7]. The study reported that:

- 36% of respondents felt that their diagnosis had been communicated sympathetically and professionally
- 40% of respondents felt that they had been given all the information necessary.

Cognitive assessments

No studies on current practice data were identified concerning the occurrence of cognitive assessment at diagnosis.

The guideline development group for the NICE 'Motor neurone disease: assessment and management' guideline considered that it is not usual practice to systematically assess cognitive function.

[6] Improving MND Care (2013) MND Association
[7] Patients' experiences of health and social care in long-term neurological conditions in England: a cross-sectional survey (2013) Peters M, Fitzpatrick R, Doll H, Playford ED and Jenkinson C. J Health Serv Res Policy January 2013 vol. 18 no. 1 28-33

4.2 Organisation of care

4.2.1 Summary of suggestions

Multidisciplinary care

Several stakeholders commented that assessment and care should be provided by a multidisciplinary team, highlighting that this improves quality of care and increases length of life. The complex nature of MND means that many different professionals need to be involved in providing care – in order to be effective multidisciplinary care therefore needs to be well coordinated.

Stakeholders highlighted how important continuity of care is to patient experience. Dealing with new carers was identified as difficult and confusing for people with MND and their families (for example, because they need to repeatedly explain their concerns and problems). A stakeholder highlighted that many professionals close cases once they have provided input – with new case workers taking over in the event of future problems. Stakeholders commented that care should be coordinated using a clinic-based multidisciplinary team.

Stakeholders highlighted the importance of carrying out cognitive assessment for people with MND, particularly as the disabling effects of MND (including communication impairment) can make it difficult to identify cognitive changes. A stakeholder also commented that the drooling of saliva (sialorrhoea) can be a problem.

Social care support

Stakeholders highlighted the importance of access to social, as well as medical, care. MND can impede an individual's ability to manage their daily living, therefore social care is a vital element of support.

4.2.2 Selected recommendations from development source

Table 5 below highlights recommendations that have been provisionally selected from the development source(s) that may support potential statement development. These are presented in full after table 5 to help inform the Committee's discussion.

Table 5 Specific areas for quality improvement

Suggested quality improvement area	Selected source guidance recommendations
Multidisciplinary care	**Organisation of care** Motor Neurone Disease. NICE clinical guideline in development. Recommendation 1.5.1 Recommendation 1.5.2 Recommendation 1.5.3
Social care support	**Information and support at diagnosis** Motor Neurone Disease. NICE clinical guideline in development. Recommendation 1.2.7 **Organisation of care** Motor Neurone Disease. NICE clinical guideline in development. Recommendation 1.5.3 **Psychological and social care support** Motor Neurone Disease. NICE clinical guideline in development. Recommendation 1.6.5

Multidisciplinary care

Motor Neurone Disease. NICE clinical guideline in development - Recommendation 1.5.1

1.5.1 Provide coordinated care for people with MND, using a clinic-based, multidisciplinary team approach.

Motor Neurone Disease. NICE clinical guideline in development - Recommendation 1.5.2

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.

Motor Neurone Disease. NICE clinical guideline in development - Recommendation 1.5.3

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).

Social care support

Motor Neurone Disease. NICE clinical guideline in development - Recommendation 1.2.7

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.

Motor Neurone Disease. NICE clinical guideline in development - Recommendation 1.5.3

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).

Motor Neurone Disease. NICE clinical guideline in development - Recommendation 1.6.5

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.

4.2.3 Current UK practice

Multidisciplinary care

No studies on current practice data were identified concerning the proportion of people diagnosed with MND who are offered multidisciplinary care. A stakeholder commented that approximately 30% of people did not have access to specialist care through MDT clinics.

The 2013 MND Association's 'Improving MND Care' survey[8] reported that 18% of respondents disagreed, or disagreed strongly, with the statement that all the different services involved in their care are coordinated well (59% of respondents agreed or agreed strongly).

A survey of patient members of the MND Association conducted between October 2008 and January 2009[9] reported that:

- almost 20% reported not having a specialist review in the last year
- almost 50% of respondents felt there was not good collaboration between health and social care services in planning care

[8] Improving MND Care (2013) MND Association
[9] Patients' experiences of health and social care in long-term neurological conditions in England: a cross-sectional survey (2013) Peters M, Fitzpatrick R, Doll H, Playford ED and Jenkinson C. J Health Serv Res Policy January 2013 vol. 18 no. 1 28-33

Cognitive assessments

The guideline development group for the NICE 'Motor neurone disease: assessment and management' guideline considered that it is not usual practice to systematically assess cognitive function.

No studies on current practice data were identified regarding the extent of cognitive assessment provided to people with MND.

Social care

The MND Association's 2013 'Improving MND care' survey reported that 13% of respondents either disagreed or disagreed strongly with the statement that they get the social care services they need when they need them (61% respondents agreed or agreed strongly with this statement)[10].

In addition, this survey also asked people to rate the care they received from the social services as a whole, responses were:

- Excellent (19%)
- Good (33%)
- Average (21%)
- Poor (8%)
- Very poor (5%)

This was lower satisfaction levels than received for the same questioned asked about NHS services (80% excellent or good responses).

[10] Improving MND Care (2013) MND Association

4.3 Respiratory impairment and cough effectiveness

4.3.1 Summary of suggestions

Assessment of respiratory function

A stakeholder highlighted the importance of assessing respiratory function, commenting that this should be done at, or shortly after, diagnosis and then monitored routinely afterwards. The stakeholder commented that onset of respiratory muscle weakness is silent and that this is often unrecognised or under-estimated by the attending physician. As the window of time between recognition of respiratory symptoms and death can be very short, it is important to introduce and establish effective therapy as soon as possible.

Cough effectiveness

Stakeholders highlighted that people with MND can have an impaired cough as a result of respiratory muscle weakness. This can lead to ineffective airway clearance and respiratory tract infection (a common cause of morbidity and mortality in this patient group). It is therefore important to monitor cough effectiveness and also to teach cough augmentation techniques. Stakeholders commented on the importance of having access to respiratory services able to provide support with these techniques.

A stakeholder also highlighted lung volume recruitment (breath stacking) as a technique that should be considered; highlighting that compared to assessment for, and provision of, non-invasive ventilation (NIV), there has been less focus on lung volume recruitment and cough assist techniques.

Provision of non-invasive ventilation (NIV)

Stakeholders commented that timely provision of NIV improves quality of life and survival. A stakeholder commented that there has been an increase in the use of NIV; however, initiation of this intervention still falls below optimum levels.

A stakeholder commented that uncontrolled oxygen should not be used prior to considering NIV (and techniques such as volume recruitment), highlighting that this can cause life-threatening carbon dioxide retention. The stakeholder also commented that in the terminal phase use of uncontrolled oxygen to provide palliation of distressing symptoms of breathlessness (often with opiates) is appropriate.

4.3.2 Selected recommendations from development source

Table 6 below highlights recommendations that have been provisionally selected from the development source(s) that may support potential statement development. These are presented in full after table 6 to help inform the Committee's discussion.

Table 6 Specific areas for quality improvement

Suggested quality improvement area	Selected source guidance recommendations
Assessment of respiratory function	**Organisation of care** Motor Neurone Disease. NICE clinical guideline in development. Recommendation 1.5.3 **Non-invasive ventilation** Motor Neurone Disease. NICE clinical guideline in development. Recommendation 1.14.8 Recommendation 1.14.10
Cough effectiveness	**Organisation of care** Motor Neurone Disease. NICE clinical guideline in development. Recommendation 1.5.3 **Cough effectiveness** Motor Neurone Disease. NICE clinical guideline in development. Recommendation 1.12.1 Recommendation 1.12.2
Provision of non-invasive ventilation (NIV)	**Respiratory impairment** Motor Neurone Disease. NICE clinical guideline in development. Recommendation 1.13.1 Recommendation 1.13.2

Assessment of respiratory function

Motor Neurone Disease. NICE clinical guideline in development - Recommendation 1.5.3

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).

Motor Neurone Disease. NICE clinical guideline in development - Recommendation 1.14.8

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 (SpO2):
    - this should be a single measurement of SpO2 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)
- sniff nasal inspiratory pressure (SNIP) and/or maximal inspiratory pressure (MIP).

Motor Neurone Disease. NICE clinical guideline in development - Recommendation 1.14.10

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.

Cough effectiveness

Motor Neurone Disease. NICE clinical guideline in development - Recommendation 1.12.1

1.12.1 Offer cough augmentation techniques to people with MND who cannot cough effectively.

Motor Neurone Disease. NICE clinical guideline in development - Recommendation 1.12.2

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

Provision of non-invasive ventilation (NIV)

Motor Neurone Disease. NICE clinical guideline in development - Recommendation 1.13.1

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.

Motor Neurone Disease. NICE clinical guideline in development - Recommendation 1.13.2

1.13.2 Offer non-invasive ventilation as treatment for people with respiratory impairment (see section 1.14).

4.3.3 Current UK practice

Assessment of respiratory function

A postal study conducted in 2009 reported on UK practice regarding the clinical application of non-invasive ventilation (NIV) in MND[11]. The study was conducted among consultant neurologists, of which 166 provided completed questionnaires. Of the responding neurologists, 38% assessed respiratory function at presentation and 20% assessed at every clinic visit. In addition, 46% of responding neurologists performed respiratory tests only if patients were symptomatic.

The MND Association's 2013 'Improving MND care' survey [12] reported that breathing assessment and monitoring occurred:

- Every few weeks (8% of respondents)
- Every few months (48% of respondents)
- An annual occurrence (11% of respondents)
- Only once (10% of respondents)
- Breathing was not monitored or assessed (19% of respondents).

[11] Non-invasive ventilation in motor neuron disease: an update of current UK practice (2012) O'Neill, CL et al. J Neurol Neurosurg Psychiatry 2012;83:371-376
[12] Improving MND Care (2013) MND Association

In addition, when asked if they ever had any difficulty in obtaining help with their breathing, 76% replied that this was 'rarely or never' the case, 8% replied that this occurred 'a few times' and 2% experienced difficulties 'regularly'.

Cough effectiveness

No studies on current practice data concerning assessment for cough effectiveness or the provision of cough augmentation techniques were identified.

Provision of non-invasive ventilation (NIV)

A postal survey conducted among consultant neurologists in the UK in 2009 reported on criteria used for referral for NIV[13]. Over three quarters of responding neurologists used selection criteria for referral for NIV: 32% used symptoms alone, 43% used symptoms and physical impairment and 10% used early intervention (patients with physiological impairment but minimal or no symptoms). The study authors commented that reliance on symptoms alone is not a sensitive way to recognise insufficient respiratory muscle weakness.

The same study also reported on the use of NIV. Of the responding neurologists, 44% had access to local NIV services and 55% had access to a regional NIV service. 1% indicated that they had no access to NIV services (this has decreased from a reported 10.2% in a survey conducted by the authors in 2000[14]).

The study authors compared results from this study with a study they had conducted in 2000. They noted that, while the number of patients seen in 2009 (1695) was very similar to 2000 (1719), that there has been a 2.6 fold increase in the number of patients referred for NIV (from 234 to 612). In addition, the number of patients currently being treated with NIV has also increased (3.4 fold; from 126 to 431). The authors concluded that, as the incidence of MND has remained stable over this time period, that this reflects changes in clinical practice.

The study authors also commented that heterogeneity in referrals made by individual neurologists exists; reporting that 7 neurologists made more than a third of the referrals for NIV and that three of these accounted for 20% of the total number. The study authors also compared the practice of the 7 highest referring neurologists to the other respondents, and reported that the frequent referrers saw more patients (50.6 compared to 8.4 mean new patients per year), monitored respiratory function more frequently and were more likely to consider early intervention with NIV.

[13] Non-invasive ventilation in motor neuron disease: an update of current UK practice (2012) O'Neill, CL et al. J Neurol Neurosurg Psychiatry 2012;83:371-376
[14] Non-invasive ventilation in motor neuron disease: current UK practice (2002) Amyotrophic Lateral Sclerosis and Other Motor Neuron Disorders 3, 145-149.

4.4 Planning for end of life

4.4.1 Summary of suggestions

A stakeholder commented that advanced care planning should occur early in disease progression because cognition and communication abilities may deteriorate as the condition progresses. Earlier planning ensures that families and professionals can be made aware of a person's wishes for end of life care before communicative or cognitive changes make this difficult or impossible. However, there is often a reluctance to discuss advance care planning (among patients, their families and healthcare practitioners).

A stakeholder also highlighted that there should be involvement of specialist palliative care from early in MND progression, commenting that palliative care improves quality of life and may extend life. They also commented that access to palliative care is variable and services may be reluctant to be involved in the care of people with MND on a longer term basis. Patients and families may also be reluctant to discuss palliative care as it is associated with end of life.

4.4.2 Selected recommendations from development source

Table 7 below highlights recommendations that have been provisionally selected from the development source(s) that may support potential statement development. These are presented in full after table 7 to help inform the Committee's discussion.

Table 7 Specific areas for quality improvement

Suggested quality improvement area	Selected source guidance recommendations
Planning for end of life	**Planning for end of life** Motor Neurone Disease. NICE clinical guideline in development. Recommendation 1.7.1 Recommendation 1.7.3 Recommendation 1.7.4

Planning for end of life

Motor Neurone Disease. NICE clinical guideline in development - Recommendation 1.7.1

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.

Motor Neurone Disease. NICE clinical guideline in development - Recommendation 1.7.3

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.

Motor Neurone Disease. NICE clinical guideline in development - Recommendation 1.7.4

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.

4.4.3 Current UK practice

The MND Association's 2013 'Improving MND Care' survey asked people with MND to identify 3 areas (out of a possible 17) that they felt should be addressed or improved as a priority[15]. 'Support with planning for the future, including later stages of MND' was ranked the third most important area; identified by 26% of respondents. Support to access palliative care and hospice services was identified by 12% of respondents.

This survey also reported that, in terms of information offered about choices and options available at the end of life:

- 43% of respondents had been offered this information
- 25% of respondents hadn't been offered information but would have liked to have and 32% of respondents didn't receive information but did not want or need it.

[15] Improving MND Care (2013) MND Association

In addition (also from the MND Association's survey results) 25% of respondents replied that they had not been given the opportunity to discuss end of life issues with a care professional. A third of respondents had replied that they felt it too early for them to have this discussion. Discussions about end of life issues were initiated by the person with MND (41%), a care professional (47%) or a family member/carer (7%).

11% of respondents commented that they had not made an Advance Care Plan or Advance Directive/Decision but would like to and 26% of respondents did not know what these were.

A postal study conducted among consultant neurologists in the UK in 2009 reported that 75% of the responding neurologists responded that they accessed specialist palliative care services at the end of life, and 69% reported that they referred to these services at an earlier stage of MND[16].

[16] Non-invasive ventilation in motor neuron disease: an update of current UK practice (2012) O'Neill, CL et al. J Neurol Neurosurg Psychiatry 2012;83:371-376

4.5 Provision of equipment

4.5.1 Summary of suggestions

Equipment and adaptations to aid activities of daily living and mobility

Several stakeholders highlighted how important it is to provide assistive equipment to people with MND. Delays in the provision of such equipment can result in people and their families struggling to manage daily activities and also reduces a person's functional independence.

Stakeholders also noted that the rapid progression of this condition means that regular reviews and reassessment of needs are required to ensure that the right equipment is provided. Ineffective provision of equipment was identified as a cause of distress to patients and their families.

In particular, stakeholders identified the provision of communication equipment and wheelchairs as areas for quality improvement.

Augmentative and Alternative Communication (AAC)

A stakeholder highlighted that most people with MND will find their voice impaired at some point. This can have a severe effect on their quality of life by reducing basic and essential interactions with others; for example, by limiting their ability to direct their care and make their wishes known. The stakeholder commented that the provision of AAC in England is being established; however, this process has encountered difficulties and progress has been slow.

Wheelchairs

A stakeholder commented that wheelchair service provision varies widely across the country, commenting that some area have fast track provision while others have rigid protocols to prevent this.

Distress/crisis interventions

A stakeholder identified the availability of the MND Just In Case kit (JIC kit) – which contains medication for the relief of anxiety and breathlessness – as an important area for quality improvement.

4.5.2 Selected recommendations from development source

Table 8 below highlights recommendations that have been provisionally selected from the development source(s) that may support potential statement development. These are presented in full after table 8 to help inform the Committee's discussion.

Table 8 Specific areas for quality improvement

Suggested quality improvement area	Selected source guidance recommendations
Equipment and adaptations to aid activities of daily living and mobility	**Organisation of care** Motor Neurone Disease. NICE clinical guideline in development. Recommendation 1.5.3 **Equipment and adaptations to aid activities of daily living and mobility** Motor Neurone Disease. NICE clinical guideline in development. Recommendation 1.9.1 Recommendation 1.9.3 Recommendation 1.9.6
Augmentative and Alternative Communication (AAC)	**Communication** Motor Neurone Disease. NICE clinical guideline in development. Recommendation 1.11.2
Wheelchairs	**Equipment and adaptations to aid activities of daily living and mobility** Motor Neurone Disease. NICE clinical guideline in development. Recommendation 1.9.4
Distress/crisis interventions	No recommendations identified.

Equipment and adaptations to aid activities of daily living and mobility

Motor Neurone Disease. NICE clinical guideline in development - Recommendation 1.5.3

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).

Motor Neurone Disease. NICE clinical guideline in development - Recommendation 1.9.1

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.

Motor Neurone Disease. NICE clinical guideline in development - Recommendation 1.9.3

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.

Motor Neurone Disease. NICE clinical guideline in development - Recommendation 1.9.6

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.

Augmentative and Alternative Communication (AAC)

Motor Neurone Disease. NICE clinical guideline in development - Recommendation 1.11.2

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 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.

Wheelchairs

Motor Neurone Disease. NICE clinical guideline in development - Recommendation 1.9.4

1.9.4 Refer people to wheelchair services without delay if needed. Wheelchair needs should be assessed and a wheelchair provided without delay.

4.5.3 Current UK practice

Equipment and adaptations to aid activities of daily living and mobility

The MND Association's 2013 'Improving MND Care' survey asked people with MND to identify 3 areas (out of a possible 17) that they felt should be addressed or improved as a priority[17]. 'Support to access specialist equipment and/or home adaptions' was ranked the fourth most important area; identified by 25% of respondents.

A cross sectional postal survey conducted between October 2008 and January 2009 gathered information on the experiences of MND carers. Of the responding carers, 20% reported that they did not receive all the necessary equipment from health and social services[18].

A survey of people with MND (conducted between October 2008 and January 2009; 505 respondents included in study) reported that 28.1% respondents reported a problem with obtaining equipment. Powered wheelchairs and ramps/rails outside the house were noted as being difficult to obtain[19].

Augmentative and Alternative Communication (AAC)

The All-Party Parliamentary Group (APPG) on Motor Neurone Disease's 'Condemned to silence'[20] reported on their inquiry into access to communication support for people with MND.

The Group noted that the NHS funds communication aids for less than half the people with MND who need them. The report authors also commented that "*Evidence we received showed that in too many areas services are poor. The*

[17] Improving MND Care (2013) MND Association
[18] Carer quality of life and experiences of health services: a cross-sectional survey across three neurological conditions (2013) Peters M, Jenkinson C, Doll H et al. Health & Quality of Life Outcomes 11: 103.
[19] Patients' experiences of health and social care in long-term neurological conditions in England: a cross-sectional survey (2013) Peters M, Fitzpatrick R, Doll H, Playford ED and Jenkinson C. J Health Serv Res Policy January 2013 vol. 18 no. 1 28-33
[20] Condemned to silence: inquiry into access to communication support for people with MND (2015) The All-Party Parliamentary Group on Motor Neurone Disease

promise of a consistent service specification for the whole country is not being delivered."

The report also contained summaries of the responses of health and social care professionals to questions about the states of services in the UK overall, including:

- In your opinion, how would you rate the level of communication support offered to people with MND?
    - 7% Excellent, 38% Good, 39% Average, 15% Poor, 2% Very Poor
- Do you feel statutory services should provide more communication support for people with MND?
    - 77% Yes, definitely, 21% Yes, to some extent, 2% No

Responses to survey questions asked of people with MND were also reported, including:

- After you were referred, how long did you have to wait for the SLT (speech and language therapist) appointment?
    - 8% A week or less, 28% 1 to 3 weeks, 39% 3 to 6 weeks, 13% 6 to 12 weeks, 5% 12 to 18 weeks, 1% more than 18 weeks.
- From the time you were told you needed communication equipment, how long did it take to receive it?
    - 7% Less than 1 week, 27% 1 to 3 weeks, 27% 3 to 6 weeks, 23% 6 to 12 weeks, 5% 12 to 18 weeks, 11% more than 18 weeks.

In addition, 20% of respondents who received communication equipment responded that it did not meet their needs and 35% of respondents were not regularly reviewed to check that equipment still met their needs.

Wheelchairs

The MND Association's 2013 'Improving MND Care' survey[21] reported that 55 % of manual wheelchair users and 71% of powered wheelchair users surveyed responded that theirs 'definitely' met their needs.

No studies on current practice data were identified concerning the provision of wheelchairs to people with MND identified as requiring them.

Distress/crisis interventions

No studies on current practice data were identified concerning the provision of MND Just In Case (JIC) kits.

[21] Improving MND Care (2013) MND Association

4.6 Additional areas

Summary of suggestions

The improvement areas below were suggested as part of the stakeholder engagement exercise. However they were felt to be either unsuitable for development as quality statements, outside the remit of this particular quality standard referral or require further discussion by the Committee to establish potential for statement development.

There will be an opportunity for the QSAC to discuss these areas at the end of the session on 26 November 2015.

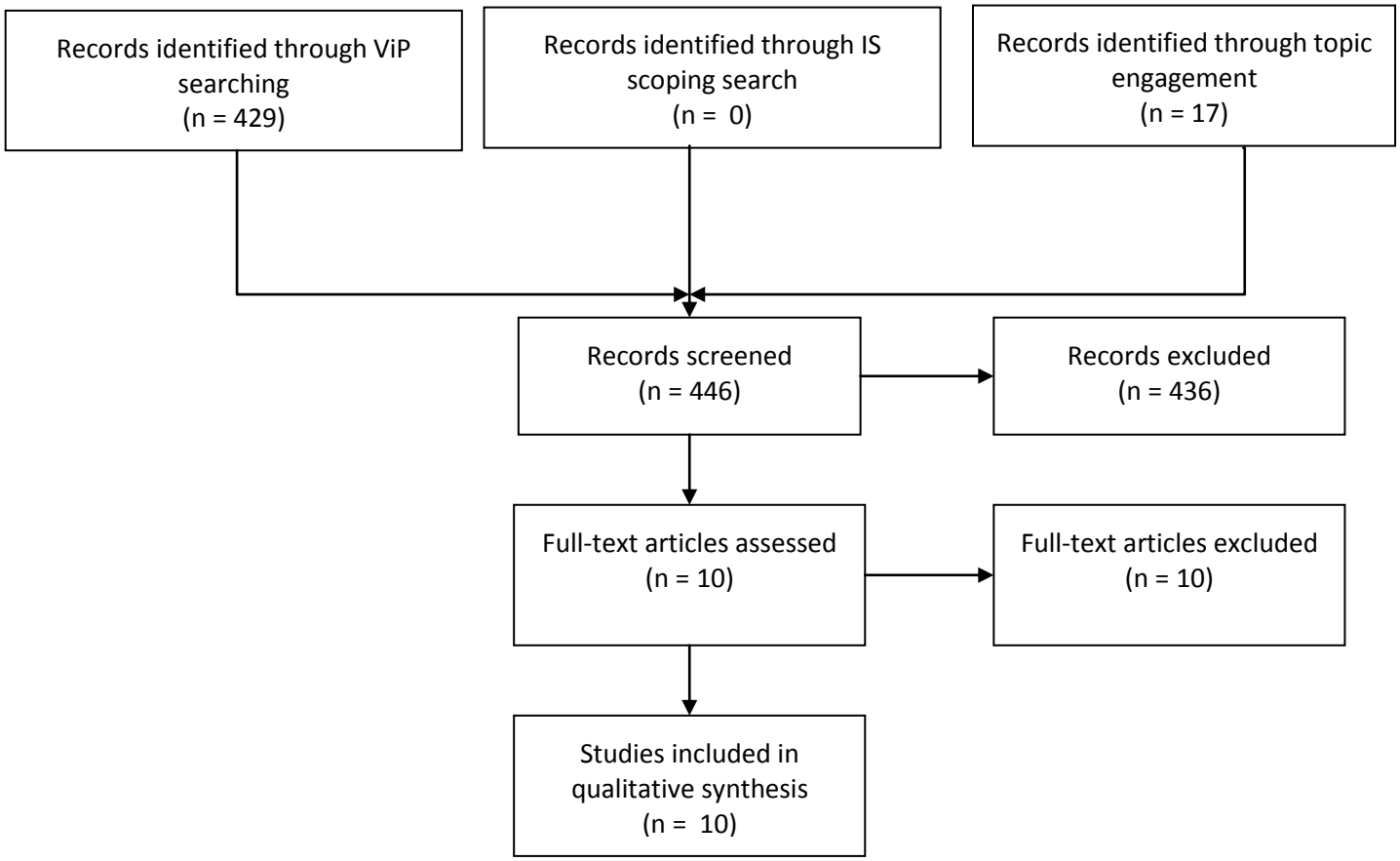
Riluzole

A stakeholder commented that people with MND should be offered riluzole (a drug that can extend life or the time to mechanical ventilation for individuals with ALS) as detailed in a NICE technology appraisal. A stakeholder also highlighted that swallowing difficulties caused by MND may make taking this drug in tablet form problematic and highlighted that suspension forms of riluzole have been developed. The stakeholder suggested that further guidance from NICE on the desirability of prescribing this form of this drug would help to prevent barriers to prescription. However, quality standards cannot use technology appraisals as basis for statements and the production of further guidance is outside the scope of quality standards.

Genetic counselling

A stakeholder suggested that genetic testing and counselling should be available to people with first or second degree relatives affected by MND, commenting that people are increasingly aware of the potential genetic basis of MND and want to know their genetic status. However, no recommendations were identified to form the basis of such a statement.

Appendix 1: Review flowchart



Appendix 2: Suggestions from stakeholder engagement exercise – registered stakeholders

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
Diagnosis					
Recognition and referral and Information and support at diagnosis					
1	Motor Neurone Disease Association	Obtaining a diagnosis of MND	The journey to a diagnosis of MND can be a highly difficult one: particular challenges include securing appropriate investigation of one's symptoms, and the way in which the final result is communicated. MND is a rare disease for which there is no diagnostic test: early symptoms could potentially be explained by other diseases or health conditions, and even once MND is suspected it must be tested for by elimination. This process can often be slow, which in turn delays treatment. Securing referral from a GP to a neurologist can often be the first major hurdle: we know that some people with MND get referred to other specialists, such as ear nose and throat or physiotherapy. Once the appropriate investigation has been carried	The Improving MND Care survey continues to identify that diagnosis remains slow: 66% of respondents replied that it was not entirely as soon as they would have liked; 52% of people were referred to a practitioner in another discipline before seeing a neurologist. The survey found that 65% of people with MND had not been advised to bring a friend or relative. We believe this is likely to be a strong indicator of a wider set of shortcomings in terms of how the diagnosis is given: we are told by people with MND that it is not always given in a private room, and that they can be left without any opportunity to gather their thoughts and compose themselves before having to travel home.	The full MND guideline examines available evidence and makes clear recommendations on best practice in referral. The MND Association's Red Flags tool, accredited by the Royal College of General Practitioners, provides a practical guide to GPs in recognising possible symptoms of MND. In respect of giving a diagnosis, the full draft guideline on MND signposts the reader to NICE Clinical Guideline 138: Patient Experiences in Adult NHS Services. In our comments on the draft guideline, we recommend that some of the key recommendations stated in CH138 should be repeated in both the short

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
			out, the end result is a piece of news as distressing as anyone is ever likely to receive. The manner in which the news is given can, however, make a substantial difference to the experience of receiving it: for too many people, the news is still broken in an inconsiderate way.		and full versions, as well as some considerations particularly relevant to MND. In particular: - In suspected cases of MND, patients should be invited to bring someone with them to all appointments, not just to the one where diagnosis is given - The diagnosis must be given in private After diagnosis, people should be given time to compose themselves in a private room.
2	Royal College of Pathologists	Key area for quality improvement 1	MND patients and their relatives and loved ones should have the opportunity to participate in research.	MND is rapidly fatal, but currently lacks an effective treatment that extends life by a reasonable amount. The leading treatment, riluzole, prolongs survival by about 2 to 3 months only1. In addition to new treatments, it is vital to develop biomarkers for diagnosis, subtyping and monitoring progress of disease. Such biomarkers may be radiological, or may be tissue- or body fluid- based2. These vital advances can only occur with the participation of patients and appropriate 'control' subjects in research.	1Miller et al. Riluzole for amyotrophic lateral sclerosis (ALS)/motor neuron disease (MND). Cochrane Database Syst Rev. 2012 Mar 14;3:CD001447. doi: 10.1002/14651858.CD001447.pub3. 2Turner MR et al. Controversies and priorities in amyotrophic lateral sclerosis. Lancet Neurol 2013; 12: 310–22

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
3	Royal College of Pathologists	Key area for quality improvement 2	Diagnostic pathways need to be robustly put in place to expedite diagnosis. More rapid diagnosis may be possible with the availability of biomarkers.	The diagnostic delay from symptom onset to formal diagnosis by a neurologist is approximately 1 year[3].	[3]Mitchell JD, et al. Timelines in the diagnostic evaluation of people with suspected amyotrophic lateral sclerosis (ALS)/motor neuron disease (MND)-a 20-year review: can we do better? Amyotroph Lateral Scler 2010;11:537–41.
4	SCM5	Key area for quality improvement 1 Information and support at diagnosis	Most GPs and health professionals have little or no knowledge of MND. Getting the right information and support at diagnosis is important both for the person and their family as they deal with this untreatable and fatal condition and enables them to access all the necessary care in a timely and appropriate manner.It needs a specialist neurologist with significant knowledge and experience of MND to lead this process	Without key information and support , the person with MND can face a traumatic and difficult disease progression , adverse effects on quality of life and significant costs to the NHS due to secondary consequences, especially when health professionals have little or no knowledge of the condition	NICE MND draft guideline
Cognitive assessment					
5	British Society of Rehabilitation	Cognition has only recently been appreciated as a problem with this group of patients and I suspect that there is poor and variable assessment of this problem nationally.			

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
	Medicine				
6	Motor Neurone Disease Association	Assessment and care of people with MND who undergo cognitive change	The link between cognitive change and MND is increasingly being understood, in respect of both the numbers of people with MND who experience cognitive change, and the biological links between some cognitive changes, including fronto-temporal dementia (FTD), and some motor disorders.	Growing understanding and awareness create new possibilities in respect of care for people with MND whose needs might not previously have been fully understood. In particular, the disabling effects of MND, including communication impairments, can make it hard to identify cognitive change. Increasing the ability of care services to overcome this barrier will bring benefits for people with MND. Securing this benefit will require greater knowledge among professionals and effective transition from assessment to care, including clarity around the responsibilities of mental health and neurology services. Currently, services commonly struggle with all of these aspects of care.	Understanding of cognitive change in relation to MND is growing quickly, and the draft guideline contains an overview. Further research is available, such as this paper on assessment: http://www.ncbi.nlm.nih.gov/pubmed/23518330.
7	SCM4	Additional developmental areas of emergent practice	Cognitive assessment of people with MND There is a research question on this in the Guidance and this will become more of an issue over the coming years	It is important to know if people with MND have / may have cognitive change as this will alter the way in which symptoms are managed and new interventions / equipment are used. If there is cognitive change there may need to be clearer discussion and careful assessment of the person's ability to benefit from an intervention / equipment.	*No additional information was provided by the stakeholder.*
8	SCM5	Additional developmental areas of emergent practice Cognitive function	Recently recognised as affecting more people with MND than previously thought		*No additional information was provided by the stakeholder.*

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
		Palliative care	Can help person with MND throughout course of disease as well as at end of life		
Organisation of care					
Multidisciplinary team					
9	British Society of Rehabilitation Medicine	There should be an emphasis on coordination of care between health and social services. This should be particularly around access to Assistive Technology. This is important because local arrangements will have been changed by the bringing of this service under Specialist Commissioning and many Social Service and Health practitioners are likely to be uncertain as to how access assessment and provision of such equipment. Assistive Technology now has a low profile to the point of it not being mentioned within the scoping document itself which is a particular concern for this group of patients.			
10	British Society of Rehabilitation Medicine	Cognition has only recently been appreciated as a problem with this group of patients and I suspect that there is poor and variable assessment of this problem nationally.			
11	British Society of Rehabilitation Medicine	Drooling continues to be an intractable problem.			
12	Motor Neurone Disease Association	Coordination and multidisciplinary care	The complex nature of MND requires the involvement of many different professionals in an individual's care. In order to be effective, this multidisciplinary care must be well coordinated.	The MND Association's Improving MND Care Survey is the largest survey of people living with MND ever undertaken in the UK; when it was last run in 2013, 950 people responded. It will be run again in 2015-16, and results will be available before this Quality Standard is finalised. Coordination of care was identified in the	In our comments on the draft guideline, we strongly recommended that the organisation of multidisciplinary care, and the role of care coordination, be looked at further. It may be that the guideline will have to

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
				2013 survey as a key problem: 72% of respondents did not strongly agree that their care was well coordinated.	recommend that this issue be prioritised for further research.
13	Motor Neurone Disease Association	Assessment and care of people with MND who undergo cognitive change	The link between cognitive change and MND is increasingly being understood, in respect of both the numbers of people with MND who experience cognitive change, and the biological links between some cognitive changes, including fronto-temporal dementia (FTD), and some motor disorders.	Growing understanding and awareness create new possibilities in respect of care for people with MND whose needs might not previously have been fully understood. In particular, the disabling effects of MND, including communication impairments, can make it hard to identify cognitive change. Increasing the ability of care services to overcome this barrier will bring benefits for people with MND. Securing this benefit will require greater knowledge among professionals and effective transition from assessment to care, including clarity around the responsibilities of mental health and neurology services. Currently, services commonly struggle with all of these aspects of care.	Understanding of cognitive change in relation to MND is growing quickly, and the draft guideline contains an overview. Further research is available, such as this paper on assessment: http://www.ncbi.nlm.nih.gov/pubmed/23518330.
14	Rotherham Doncaster and South Humber NHS FT	Key area for quality improvement 2 Continuity of care	Continuity of care is where health care is provided for a person in a coordinated manner and without disruption despite involvement of different practitioners in different care settings	Continuity of care becomes increasingly important for patients as they age, develop multiple morbidities and complex problems, or become socially or psychologically vulnerable.Continuity of care contributes importantly to patient experience	http://www.kingsfund.org.uk/sites/files/kf/field/field_document/continuity-care-patient-experience-gp-inquiry-research-paper-mar11.pdf
15	SCM2	Key area for quality improvement 1 MDT Co-ordinated care	There is good evidence that coordinated care for people with MND, using a clinic-based, multidisciplinary team approach	Recent cohort studies such as Aridegbe (2013) and Rooney (2015) supports the benefit of an MND MDT based in a clinic setting but with clear arrangements in place	Please see the MND Association MDT Working Best Practice Guide www.mndassociation.org/

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
		for patients with MND	can increase in survival time and increased uptake of interventions for MDT care therefore preventing a decent into crisis and the need for NIV.	for close collaboration with care providers in the community on survival as well as being clinically and cost effective. There remains variation in how MND services are structured with some patients not having access to MND MDT service or reviews with some studies indicating that this leads to delays in accessing appropriate intervention such as NIV which can impact on quality of life and survival time.	wp-content/uploads/2015/02/MND%20MDT%20Guide%202011a.pdf
16	SCM3	Provision of MDT care through attendance multidisciplinary clinic as described in the guidance	Attendance at a specialist MDT clinic has improved survival outcomes	A large proportion of patients (approx. 30%) do not have access to specialist care through an MDT clinic	*No additional information was provided by the stakeholder.*
17	SCM4	Multidisciplinary team assessment of patient with MND	There is good evidence that multidisciplinary team care improves quality of life and may increase length of life	There are excellent MDT in the MND Care centres across the country but not all patients with MND are able to attend a centre	The Economic evaluation suggests that MDT care is cost effective
18	SCM4	Continuity of professionals and carers in the care of patients	Many patients with MND and their families find the variability in the carers involved very difficult, as they need to repeatedly explain about their problems and concerns. Many professionals close cases following an input and when there are further problems a new case worker may then take over. In social care research has shown	Within the research these areas were identified as an area of concern and distress for patients and families. This varies across the country and we should aim to replicate the best practice universally.	*No additional information was provided by the stakeholder.*

ID	Stakeholder	Suggested key area for quality improvement	Why is this important?	Why is this a key area for quality improvement?	Supporting information
			that the continual changing of carers, undertaking personal care, is confusing and difficult for patients and families – as time is taken in explaining the disease and the person's specific needs and the care provided is haphazard and ineffective.		
19	SCM 4	Additional developmental areas of emergent practice	Cognitive assessment of people with MND There is a research question on this in the Guidance and this will become more of an issue over the coming years	It is important to know if people with MND have / may have cognitive change as this will alter the way in which symptoms are managed and new interventions / equipment are used. If there is cognitive change there may need to be clearer discussion and careful assessment of the person's ability to benefit from an intervention / equipment.	*No additional information was provided by the stakeholder.*
20	SCM 5	Key area for quality improvement 3 Single point of contact and MDT care	People with MND need access to many specialist areas of health and social care. Coordinated care is vital to ensure access to the right professionals at the right time and specialist multi-disciplinary care gives best quality of care and cost effectiveness	Improves the journey with MND for the patient and their family and reduces the secondary and expensive consequences of unplanned hospital admission , inappropriate care and avoidable crises.	NICE MND draft guideline
21	SCM5	Additional developmental areas of emergent practice Cognitive function	Recently recognised as affecting more people with MND than previously thought		*No additional information was provided by the stakeholder.*

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		Palliative care	Can help person with MND throughout course of disease as well as at end of life		
Social care support					
22	Motor Neurone Disease Association	Social care	The effects of MND serve to impede the individual's ability to manage daily living, drastically by the time the disease is well advanced. Social care is therefore a vital day-to-day element of support for people with MND, and also crucial to the experience of carers.	The Improving MND Care Survey identified markedly lower satisfaction among people with MND with social care provision than with healthcare provision. There is also abundant evidence that reduced funding for local authorities, standardised but sometimes more stringent eligibility criteria and increased charges by local authorities are all combining to make high quality social care ever-more difficult to obtain, certainly with statutory funding.	The full guideline recognises the importance of social care to people with MND and carers, and outlines available research.
23	SCM 5	Key area for quality improvement 5 Social care provision for MND	People with MND need access to social as well as medical care. If social care doesn't work well for them , the on costs to the NHS can be huge and detrimental to both the health of the person and their immediate carers.	The journey with MND is complex and difficult. Getting the right provision at the right time of both health and social care impacts hugely on both quality of life , ability to function and health budgets	NICE MND draft guideline
Respiratory impairment and cough effectiveness					
Assessment of respiratory function					
24	SCM 1	Assessment of respiratory function at, or shortly after, diagnosis of	The onset of respiratory muscle weakness is silent, and if respiratory support is not	This recommendation is supported by current NICE guidelines, and the draft update for consultation. The most recent	Fallat Arch Neurol 1979;36:74-80 Lyall Brain 2000;124:2000-

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		MND. This assessment should include a) bulbar function (influences further tests – see NICE CG105/update in progress), b) respiratory muscle function, c) cough effectiveness.	provided, the window between recognition of respiratory symptoms and death may be very short, leaving little time to introduce and establish effective therapy. In most people with MND, respiratory muscle weakness is present at the time of diagnosis, but in the absence of lung function, this tends to go unrecognised, or to be under-estimated, by the attending clinician.	national survey of respiratory care in MND showed that 38% of Neurologists assessed lung function at presentation. Among the Neurologists who referred the highest number of patients for consideration of non-invasive ventilation (NIV), all assessed lung function at presentation.	13 NICE: CG105 & current draft MND guideline for consultation. JNNP 2012;83:371-76
25	SCM 1	Both respiratory symptoms, and respiratory function, should be monitored routinely. If a patient has clearly expressed a wish that they do not want to consider any form of ventilatory support, assessment may be limited to symptoms (to inform alternative palliation).	The rate of decline in lung function is highly variable. Ideally, the role of NIV and other forms of respiratory support should be discussed with patients and their family /carers in advance, rather than in a crisis situation. Regular monitoring of both symptoms and function will identify patients who may benefit from respiratory support at an earlier stage. Structured respiratory care, including routine assessment of respiratory symptoms and tests of respiratory function, is associated with better outcomes, including a higher proportion of patients receiving NIV, a lower proportion	Compared to the previous National UK survey of respiratory care in MND, the most recent survey showed substantial improvements in the use of NIV, including a 3.4 fold increase in patients receiving NIV, and a higher proportion successfully established on NIV. However there is room for further improvement. The Neurologists who referred the highest number of patients for NIV were more likely to routinely assess respiratory function at each clinic visit, and to base referral for NIV on the combination of respiratory symptoms and physiology (function), consistent with other research. However, in most clinics, respiratory function was not routinely assessed. When including assessment of respiratory function either routinely or if symptoms were present,	Ferrero Chest 2005;127:2132-8 NICE: CG105 & current draft MND guideline for consultation. JNNP 2012;83:371-76

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			initiated on ventilation in an emergency and improved survival from diagnosis (Ferrero 2005).	this was only performed by 66% of respondents.	
Cough effectiveness					
26	British Society of Rehabilitation Medicine	The provision of specialist respiratory services to patients with neurological disability is patchy and many districts will be not be providing cough assist equipment.			
27	SCM 1	In addition to NIV, lung volume recruitment and sputum clearance should be considered.	Lung volume recruitment helps reverse or prevent lung atelectasis (areas of collapse), improving gas exchange. This is also essentially the first step of various cough assist techniques, which help improve clearance of secretions from the airway. Such techniques are not infrequently indicated prior to the requirement for ventilatory support.	Compared to assessment for, and provision of, NIV, there has been less focus on both volume recruitment and cough assist techniques, although areas of excellent practice exist. In addition to equipment, such as mechanical insufflation-exsufflation devices, simple breath-stacking using an ambu-bag, one-way valve and face mask is a cheap and effective option for many patients, and should be considered first line.	NICE Management of Motor Neurone Disease: Draft for consultation (September 2015). Armstrong et al. British Journal of Nursing 2009;18:1094-7. Chatwin et al. Eur Respir J; 2003;21:502-8. Sancho et al. Chest 2004;125:1400-5.

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28	SCM 2	Key area for quality improvement 2 Regular monitoring of PCF (just like asthma patients measure their PFR) by MND patients and their carers with an action plan in place to facilitate effective cough if their PCF falls below effective levels. Access to a HCP ideally a respiratory physiotherapist to teach cough augmentation techniques when a patient's PCF fall below threshold levels with regular reviews to modify techniques as the individual's condition progresses.	An effective cough is a vital for clearing airway secretions and protecting against respiratory tract infections, which are the commonest cause of hospital admissions in patients with respiratory muscle weakness due to neuromuscular disease. Patients with MND may have an impaired cough and reduced peak cough flow as a result of respiratory muscle weakness. A reduced peak cough flow (PCF) leads to ineffective airway clearance which can accelerate the development of infection, pneumonia s and respiratory failure and as such has been identified as a major cause of morbidity and mortality in this patient group. The use of cough augmentation techniques have been shown to help secretion clearance, decrease hospitalisation rates, prevent, or delay the need for tracheostomy and prolong survival when used in conjunction with NIV.	Airway clearance techniques are aimed at helping the removal of secretions. Independent airway clearance techniques are ineffective in patients with compromised respiratory muscles. Conventional physiotherapy techniques used to assist secretion clearance such as postural drainage are unlikely to be effective in this client group, and suction is neither well tolerated nor therefore successful. Thus patients with compromised respiratory muscles require assisted coughing, non-invasive assistance and aids for airway clearance with regular monitoring. However access to assessment for and provision of these adjuncts varies. In addition their funding and availability is still limited across the UK particularly in the community setting. In some cases or areas patients descend into respiratory crisis and they are admitted to the acute setting before impaired cough and mechanism to improve its effectiveness is considered.	www.brit-thoracic.org.uk/document-library/clinical-information/physiotherapy/physiotherapy-guidelines/physiotherapy-guideline/ Section 6c and supporting appendices The MD Association Pathway http://www.mndassociation.org/wp-content/uploads/2015/04/MND-Respiratory-Pathway-v1.1-070615.pdf http://www.rcjournal.com/cpgs/pdf/nonpharmacologic_2013.pdf Vitacca M, Paneroni M, Trainini D, Bianchi L, Assoni G, Saleri M, Gile` S, Winck JC, Gonc¸alves MR: At Home and on Demand Mechanical Cough Assistance Program for Patients With Amyotrophic Lateral Sclerosis. Am J Phys

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					MedRehabil 2010;89:401–406 Bento, J. Goncalves, M. Silva, N. Pinto, T. (2010) Indications and Compliance of Home MI-E in Patients with NMD. Arch Bronconuemol. 46(6) 420-425 Expert in the field is Dr Michelle Chatwin and would be an excellent person to advise and present on this topic
Provision of non-invasive ventilation (NIV)					
29	SCM 1	Uncontrolled oxygen should not be used prior to considering NIV (and techniques such as volume recruitment), except in the palliative setting, when such interventions have been deemed inappropriate.	In patients with respiratory muscle weakness who are not receiving ventilatory support, uncontrolled oxygen therapy can cause life-threatening carbon dioxide retention, and should be avoided. However, in the terminal phase, use of uncontrolled oxygen to provide palliation of distressing symptoms of breathlessness (often with opiates) is appropriate.	The last UK survey of respiratory care in MND showed that 26% of responders used uncontrolled oxygen prior to the end of life in at least some patients with symptomatic respiratory impairment, who had not been assessed for NIV.	Gay Mayo Clin Proc 1995;70:327-30. O'Driscoll Thorax 2008;63(Suppl 6):vi1-68 – British Thoracic Society guideline for emergency oxygen use in adult patients. JNNP 2012;83:371-76
30	SCM 3	All patients with MND should be assessed for respiratory failure and	NIV is the single most effective intervention terms of improving survival and quality of life.	A survey of UK neurologists indicated an increase in use of NIV for patients with MND since the introduction of NICE guidance but	*No additional information was provided by the stakeholder.*

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		have NIV initiated when indicated		this till falls short of everyone receiving the appropriate screening and initiation of this intervention	
31	SCM 5	Key area for quality improvement 4 Provision of Non invasive ventilation	Timely provision of NIV improves quality of life for the person with MND and also improves survival.	Ensuring the service is available and set up to support the person with MND and easily accessed throughout England.	NICE MND draft guideline
Provision of equipment					
Equipment and adaptations to aid activities of daily living and mobility					
32	SCM 2	Key area for quality improvement 3 Provide appropriate equipment and adaptations without delay to maximise people's participation in activities of daily living and maintain their quality of life.	Patients and their carers find that timely access to appropriate assistive equipment to maintain functional independence and quality of life varies across the UK as is the appropriate training to use the equipment safely in some circumstances. This sometimes means inappropriate equipment is ordered for patients that does not meet their needs which has an impact on the patients maintenance of their functional independence for as long as possible and upon their psychological wellbeing and that of their carers.	Any delays in the appropriate provision of equipment can result in patients and their families struggling to safely manage activities of daily living in the home setting. Such situations can tip the patient into crisis before the shortfall in equipment provision is addressed i.e. post fall. Standards in relation to equipment provision would support a more proactive than reactive approach to maintaining the patient's functional independence. The tendency toward rapid progression of the disease necessitates regular reviews and reassessment of equipment, and makes planning for the future essential in any adaptations to the home. Liaison with home care workers is important, to ensure they are aware of the implications of the condition.	http://www.mndassociation.org/life-with-mnd/Publications-pabmnd/living-with-motor-neurone-disease-guide/
33	SCM 4	Equipment to aid daily living and facilitate communication should be	Research shows that the ineffective provision of equipment – not suitable to the patient's	The provision of the correct equipment, which is able to be adapted to cope with the disease progression, in a timely way varies	*No additional information was provided by the stakeholder.*

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		provided appropriately and in a timely manner, and allow for further adaption as the disease progresses	needs or after a long delay so that it is no longer appropriate and helpful – is an area of distress to patients and families. This applies to both equipment to aid daily living, such as wheelchair, or for communication, augmentative and alternative communication aids.	across the country. The expertise of professionals advising on equipment varies, as does their experience in managing the complex issues of MND. The timely provision of quality of life for patient and family, allows the patient to remain more independent, reduce falls an incidents that could lead to admission to hospital and deterioration.	
34	SCM 5	Key area for quality improvement 2 Timely and appropriate provision of equipment particularly wheelchairs and communication aids	Provision of equipment to aid function , mobility and communication , enables people with MND to sustain some quality of life, reduce care costs and function to the best of their ability.	Timely and appropriate provision is essential , both in terms of enabling the individual through an often rapidly changing disease whilst avoiding unnecessary costs to the NHS in terms of wasted provision	NICE MND draft guideline
Augmentative and Alternative Communication (AAC)					
35	Motor Neurone Disease Association	Augmentative and Alternative Communication (AAC)	Most people with MND will find their voice impaired at some point during their illness. This can have a devastating impact on their quality of life, both by curtailing basic and essential interaction with others, and by limiting their ability to direct their care and make their wishes known, not least at the end of life. There is much evidence that appropriate use of AAC, which can be low or high tech, can make a substantial difference to the	Provision of AAC in England, and also to some extent Wales, is undergoing a transformation: it is now clearly recognised as a health need, and clearer systems for providing it are being established. However, this process has encountered many difficulties in England particularly, and progress has been slow: a report by the All Party Parliamentary Group on MND found early in 2014 that delivery had so far been a failure. NHS England is improving the situation, but the centrality of communication to a person's experience of MND, and the rapidly changing nature of technology, means that it will require close	The APPG report 'Condemned to Silence' outlines the benefits of AAC in detail, and also sets out difficulties with NHS England's implementation up to late 2014. NHS England's service specification for specialised AAC (D01/S/b) outlines responsibilities on NHS providers.

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			person's quality of life.	attention for years to come.	NHS England has developed a minimum data set for specialised AAC, and is in the early stages of regular data collection. AAC is referenced in the draft NICE guideline on MND.
Wheelchairs					
36	SCM 3	Wheel chair provision needs to be more timely	MND has a fast trajectory and needs change quickly. Being able to anticipate patient needs and provide a wheelchair when it is required is ideal	Currently wheelchair service provision varies greatly across the country. There is inequity of provision. In some areas patients with MND are fast tracked. In other areas rigid protocols prevent this. The result is that a patient will not be seen until many months after the need for a wheelchair.	*No additional information was provided by the stakeholder.*
Distress/crisis interventions					
37	Rotherham Doncaster and South Humber NHS FT	Key area for quality improvement 1 Distress/crisis interventions	The MND Just In Case kit (JIC kit) The MND JIC kit is designed to hold medication for the relief of anxiety and breathlessness.	Its presence in the home provides tangible evidence for people with MND and carers that fears have been addressed and practical help is at hand.	http://www.mndassociation.org/wp-content/uploads/Motor-neurone-disease-a-guide-for-GPs-and-primary-care-teams-v1.1-Jul15.pdf
Planning for end of life					
38	SCM 4	**Assessment and involvement of specialist palliative**	There is good evidence that palliative care improves quality of life and symptoms, and may	Access to specialist palliative care varies across the country and there may be reluctance for services to be involved in the	There is increasing evidence for palliative care:

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		care from early in the disease progression	extend life.	care of people with MND on a longer term basis. There may also be reluctance of patients and families to discuss and accept palliative care, which is often associated with end of life.	Temel JS, Greer JA, Muzikansky et al . Early palliative care for patients with metastatic non-small-cell lung cancer. N Eng J Med 2010; 19: 733-742. Zimmermann C, Riechelmann R, Krzyzanowska M, Rodin G, Tannock I. Effectiveness of specialized palliative care: a systematic review. JAMA 2008; 299: 1698-709. Veronese S, Gallo G, Valle A, Cugno C, Chio A, Calvo A, Rivoiro C, Oliver DJ. The palliative care needs of people severely affected by neurodegenerative disorders: a qualitative study. Prog Pall care 2015; DOI: http://dx.doi.org/10.1179/1743291X15Y.0000000007
39	SCM 4	**Discussion of advance care planning early in the disease progression**	There is increasing evidence that advance care planning can be helpful in caring for people at the end of life, particularly, as in MND, when communication and	There is often reluctance to discuss advance care planning – as this may be seen to be upsetting or patients and families may not wish to look ahead to end of life. This reluctance may be with professionals,	*No additional information was provided by the stakeholder.*

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			cognition may deteriorate as the disease progresses. Families and professionals are then more aware of the patient's wishes and the patient's autonomy may be maintained.	families, patients and within society as a whole. Clearer advance care planning would allow patients to express their views, before communication and cognitive change makes this more difficult or impossible, and allow improved care, with reduced unwanted interventions or admission to hospital.	
40	SCM 5	**Additional developmental areas of emergent practice** **Cognitive function** **Palliative care**	Recently recognised as affecting more people with MND than previously thought Can help person with MND throughout course of disease as well as at end of life		*No additional information was provided by the stakeholder.*
Additional areas					
Riluzole					
41	Motor Neurone Disease Association	Additional developmental areas of emergent practice: suspension riluzole	Riluzole is the only medicine known to extend survival in MND and licensed for use on the NHS. It is covered by NICE Technology Appraisal 20. However, the swallowing difficulties commonly caused by MND make taking a medicine in tablet form problematic. Some	Two suspension forms of riluzole have been developed, by separate companies. Even in its regular tablet form, rilzuole can sometimes attract objections from GPs, who feel it is expensive and decline to prescribe it, despite the existence of the technology appraisal. There is a risk that similar barriers to obtaining the suspension forms of riluzole could be even more commonplace. Guidance from NICE to affirm the	TA20 remains an authority on riluzole. The two companies who have developed the suspension forms should be able to provide further data.

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			people crush the tablet and either take it with a liquid such as honey, or administer it via gastrostomy tube.	desirability of prescribing a suspension form when appropriate would help to prevent this.	
42	SCM 3	All patients should be offered riluzole unless contraindicated	Riluzole is the only disease modifying medication, approved by NICE.	It is important to ensure that all patients with MND are being offered this disease modifying therapy.	*No additional information was provided by the stakeholder.*
Genetic counselling					
43	Royal College of Pathologists	Key area for quality improvement 3	Genetic testing and counselling should be available to patients with first or second degree relatives affected by MND. This should be requested by a physician who: 1) understands the significance of the potential results, 2) is capable of communicating this to the patient and 3) can arrange suitable care and support after a result is made known.	Approximately 5-10% of ALS cases are familial (having one or more similarly affected first degree relative and a substantial proportion of seemingly "sporadic" disease is associated with mutations of *C9ORF72.* Patients are increasingly aware of this and the issues surrounding genetic counselling and inheritance of these disorders. Families and patients increasingly demand to know what their relevant genetic status is.	Chiò et al. Genetic counselling in ALS: facts, uncertainties and clinical suggestions. J Neurol Neurosurg Psychiatry 2014;85:478–485. doi:10.1136/jnnp-2013-305546
No comment					
44	NHS England	Thank you for the opportunity to comment on the above Quality Standard. I wish to confirm that NHS England has no substantive comments to make regarding this consultation.			
45	Royal College of Nursing	This is to inform you that the RCN has no comments to submit to inform on the above topic engagement at this time.			