UKMi North West

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A summary of prescribing recommendations from NICE guidance

Motor Neurone Disease

NICE NG42; 2016

This guideline includes recommendations on assessing and managing MND, a neurodegenerative condition affecting the brain and spinal cord. The most common type of MND is amyotrophic lateral sclerosis.

Definition of terms

MND	motor neurone disease
MDT	multidisciplinary team
U	unlicensed

Assessment and referral

- Ensure robust protocols and pathways are in place to inform healthcare professionals about MND and how it may present, local arrangements for referral and to ensure continued and integrated care for MND across all care settings.
- Be aware MND causes progressive muscular weakness that may first present as isolated and unexplained symptoms such as:
  - functional effects of muscle weakness e.g. loss of dexterity, falls or trips,
  - speech or swallowing problems, or tongue fasciculations (bulbar presentation),
  - muscle problems e.g. weakness, wasting, twitching, cramps and stiffness,
  - breathing problems e.g. shortness of breath on exertion or respiratory symptoms that are hard to explain,
  - effects of reduced respiratory function e.g. excessive daytime sleepiness, fatigue, early morning headache or shortness of breath when lying down.
- Be aware that MND may first present with cognitive features, which may include:
  - behavioural changes,
  - emotional lability (not related to dementia),
  - frontotemporal dementia.
- If MND is suspected, refer without delay and specify possible diagnosis in referral letter. Contact the consultant neurologist directly if the person needs to be seen urgently.
- Provide information and support for people, their family and/or carers throughout the diagnostic process, particularly during periods of diagnostic uncertainty or delay.

Cognitive assessment

- Be aware that people with MND and frontotemporal dementia may lack mental capacity. Care should be provided in line with the Mental Capacity Act 2005.
- Explore any cognitive or behavioural changes with the person, their family and/or carers as appropriate at diagnosis, and if there is concern about cognition and behaviour. If needed, refer for a formal assessment - see NICE pathway: Dementia.
- Tailor discussions to the person's needs, taking into account communication ability, cognitive status and mental capacity.

Organisation of care – see NICE pathway

- Provide coordinated care for people with MND, using a clinic-based (hospital or community) specialist MDT approach.
- The MDT should:
  - include healthcare professionals and social care practitioners with expertise in MND, and staff who see people in their home,
  - ensure effective communication and coordination between all healthcare professionals and social care practitioners involved in the person's care and their family members and/or carers (as appropriate),
  - carry out regular, coordinated assessments at the MDT clinic (usually every 2 to 3 months) to assess people's symptoms and needs,
  - provide coordinated care for people who cannot attend clinic, according to person's needs.

Information and support – see NICE pathway

Treatment and symptom management

Muscle problems

- Discuss available treatment options taking into account the person's needs and preferences, and whether they have any difficulties taking medicine (e.g. problems swallowing).
- ***First-line;*** consider quinine**U** for muscle cramps. If quinine is contraindicated, not tolerated or not effective consider baclofen**U.** If baclofen is contraindicated, not tolerated or not effective consider, tizanidine**U**, dantrolene**U** or gabapentin**U**.
- Consider baclofen, tizanidine, dantrolene**U** or gabapentin**U** to treat muscle stiffness, spasticity or increased tone. If not effective, not tolerated or contraindicated, consider referral to a specialist service for the treatment of severe spasticity.
- Review treatments for muscle problems during MDT assessments; ask how the person is finding treatment, whether it works and if they have any adverse effects.
- Consider an exercise programme for people with MND - see NICE pathway

Saliva problems

- Assess volume and viscosity of saliva, respiratory function, swallowing, diet, posture and oral care.
- If a person with MND has problems with drooling of saliva (sialorrhoea), provide advice on swallowing, diet, posture, positioning, oral care and suctioning.
- ***First-line*****:** consider a trial of an antimuscarinic**U β** for sialorrhoea in people with MND without cognitive impairment.
- For people with MND and cognitive impairment consider glycopyrrolate**U** for sialorrhoea as it has fewer central nervous system effects**β**.
- If first-line treatment is not effective, not tolerated or contraindicated, consider referral to a specialist service for botulinum toxin A**U**.
- If a person with MND has thick, tenacious saliva:
  - review all current medicines, especially any treatments for sialorrhoea,
  - provide advice on swallowing, diet, posture, positioning, oral care, suctioning and hydration,
  - consider treatment with humidification, nebulisers and carbocisteine.

β Editorial note: Also see the following documents:
NICE advice [ESUOM15]; Oral glycopyrronium bromide
UKMi Medicines Q&A's: Hypersalivation – what drug treatment options are available?; Hypersalivation – what are the treatment alternatives to glycopyrronium and hyoscine?

Motor neurone disease continued……

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Speech and communication – see NICE pathway

Support for Activities of Daily Living

- Healthcare professionals and social care practitioners, including physiotherapists and occupational therapists, should assess and anticipate changes in the person's daily living needs - see NICE pathway.

Respiratory function and symptoms

- Assess and monitor the person's respiratory function - see NICE pathway and symptoms - see NICE pathway.
- Treat people with MND and worsening respiratory impairment for reversible causes (e.g. respiratory tract infections or secretion problems), before considering other treatments.
- Offer non-invasive ventilation treatment for people with respiratory impairment; decisions should be made by the MDT in conjunction with the respiratory ventilation service, and the person - see NICE pathway,
- Consider urgent non-invasive ventilation for people with MND who develop worsening respiratory impairment and are not already using this.
- Consider opioids**U** as an option to relieve symptoms of breathlessness. Take into account the route of administration and acquisition cost.
- Consider benzodiazepines**U** to manage breathlessness that is exacerbated by anxiety. Take into account the route of administration and acquisition cost of medicines.
- Offer cough augmentation techniques such as manual assisted cough to people with MND who cannot cough effectively.
- Consider unassisted breath stacking and/or manual assisted cough as the first-line treatment for people with MND who have an ineffective cough.
- For people with bulbar dysfunction, or whose cough is ineffective with unassisted breath stacking, consider assisted breath stacking (e.g. using a lung volume recruitment bag).
- Consider a mechanical cough assist device if assisted breath stacking is not effective, and/or during a respiratory tract infection.

Nutrition and gastrostomy – see NICE pathway: MND and nutrition support in adults

- Assess the person's weight, diet, nutritional intake, fluid intake, hydration, oral health, feeding, drinking and swallowing, and offer support, advice and interventions as needed at diagnosis, at MDT assessments, or if there are any concerns about weight, nutrition or swallowing.
- If swallowing problems are suspected arrange a clinical swallowing assessment.
- Discuss gastrostomy at an early stage, and at regular intervals as MND progresses, taking into account the person's preferences and issues, such as ability to swallow, weight loss, respiratory function, effort of feeding and drinking and risk of choking. Be aware that some people will not want to have a gastrostomy.
- Explain the benefits of early placement of a gastrostomy, and possible risks of a late gastrostomy (e.g. low critical body mass, respiratory complications, risk of dehydration, different methods of insertion, and a higher risk of mortality and procedural complications).
- If a person is referred for a gastrostomy, it should take place without unnecessary delay.

Riluzole for amyotrophic lateral sclerosis MND

See NICE TA20

- Riluzole is recommended for treatment of individuals with amyotrophic lateral sclerosis MND.
- Therapy should only be started by a neurological specialist experienced in management of MND.
- Routine supervision of therapy may be managed by GPs acting under a shared care agreement.

Psychological and social care – see NICE pathway

- During MDT assessments and other appointments, discuss the psychological and emotional impact of MND with the person, their family and/or carers and ask whether they have any psychological or support care needs.
- Offer the person information about sources of emotional and psychological support, including support groups and online forums. If needed, refer the person to counselling or psychology services for a specialist assessment and support.
- Offer family and/or carers information about respite care and sources of emotional and psychological support, including support groups, online forums and counselling or psychology services.
- Be aware that as MND progresses, people may develop communication problems and have difficulty accessing support or services e.g. they may be unable to access a call centre). Ensure people are given different ways of getting in touch with support or services, and a designated contact if possible.

End of life care – see NICE pathway

- Offer opportunities to discuss preferences and concerns about care at the end of life at trigger points such as: at diagnosis, if there is a significant change in respiratory function, or if interventions such as gastrostomy or non-invasive ventilation are needed. Be sensitive about the timing of discussions and take into account the person's current communication ability, cognitive status and mental capacity.
- Be prepared to discuss end of life issues whenever people wish to do so.
- Provide support and advice on advanced care planning for end of life and consider discussing this at an earlier opportunity if it is expected that their communication ability, cognitive status or mental capacity will get worse.
- Provide additional support as the end of life approaches e.g. additional social or nursing care to enable carers and family to reduce their carer responsibilities and spend time with the person with MND.
- Towards the end of life, ensure there is prompt access to the following, if not already provided:
  - a method of communication that meets the person's needs,
  - specialist palliative care,
  - equipment, if needed, syringe drivers, suction machines, riser–recliner chair, hospital bed, commode and hoist,
  - anticipatory medicines, including opioids and benzodiazepines to treat breathlessness, and antimuscarinic medicines to treat problematic saliva and respiratory secretions.
- Offer bereavement support to family and/or carers.

Recommendations – wording used such as 'offer' and 'consider' denote the strength of the recommendation.

Drug recommendations – the guideline assumes that prescribers will use a drug's Summary of Product Characteristics (SPC) to inform treatment decisions.

This bulletin summarises key prescribing points from NICE guidance. Please refer to the full guidance at www.nice.org.uk for further detail. This is an NHS document not to be used for commercial purposes.