Motor neurone disease: a review of the new NICE guideline

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The authors review the guideline on motor neurone disease published earlier in 2016 by the National Institute for Health and Care Excellence (NICE). They welcome the focus on symptomatic management and the use of data extrapolated from interventions in other neurological conditions, noting that ‘such a common sense approach is particularly helpful in these times of austerity’.

In 2005, after merging with the health development agency, NICE changed its name from the National Institute for Clinical Excellence to the National Institute for Health and Clinical Excellence. This minor change in name coincided with a significant move from its original remit. When it was originally set up in 1999, NICE’s remit was: to reduce the variation in the availability and quality of NHS treatments and care. The current, more general, brief is: developing public health guidance to prevent illness and promote health. Legislation in 2013 widened its responsibility further to include producing social care standards relevant to health issues, and a further name change was made to the National Institute for Health and Care Excellence.

This journey of evolution and change is mirrored by motor neurone disease (MND) NICE guideline development. The guideline for the use of riluzole in MND was published in 2001 to deal with a single clinical intervention, carrying out a thorough analysis of its effectiveness and eventually giving a clear conclusion regarding its cost effectiveness. The 2010 guidance on the use of non-invasive ventilation in patients with MND took a further step to give a more practical and pragmatic approach to the clinical intervention evaluated. The February 2016 guideline for the assessment and management of MND read more like royal college or specialist society guidelines with more reliance on consensus and expert opinion than on robust evidence from controlled trials.

NICE was clear that the guideline’s main aim is to offer best practice recommendations on multiple issues including communication of diagnosis, monitoring of disease progression, symptom management and preparation for end-of-life care. This should lead to standardised and improved care for patients with MND, focussing on a multidisciplinary team (MDT) approach, with attention to patient concerns and quick response to any deterioration.

Key recommendations

Diagnosis and prognosis

The recommendations recognise that early referral is important, enabling patients to receive early assessment, investigations and eventually, accurate diagnosis. MND leads to progressive muscular weakness. However, it can present with isolated and unexplained symptoms such as muscle weakness leading to falls, loss of dexterity, bulbar symptoms or symptoms of reduced respiratory function. Cognitive impairment was stressed as an unusual but important presenting feature. The accumulating evidence linking MND with frontotemporal dementia validates such recommendation. The guideline highlights the recently developed Red Flag diagnosis tool developed by the MND Association and RCGP.

The guidelines recommend that the diagnosis should be given by an expert neurologist, so that all aspects of care can be discussed. There should be a follow-up appointment within four weeks with a designated healthcare professional; currently this is likely to be an MND specialist nurse. Throughout the guideline, the emphasis on patient wishes and family involvement and support is emphasised.

As part of the initial assessment the guidance recommends performing an assessment of cognitive symptoms and behavioural changes (expert opinion). This guidance does not recommend a particular screening tool as there are no validated tools used specifically in MND.

When considering prognosis, only one externally validated tool for predicting survival was found – the amyotrophic lateral sclerosis (ALS) Prognostic Index. However, the research using this tool was observational in nature and deemed low-quality evidence. The guideline offers a list of prognostic factors. These include bulbar presentation, weight loss, poor respiratory function, older age and a shorter time from first symptoms to diagnosis. A low ALS function rating scale at baseline is also included as an indication for poor prognosis.
Management
An ongoing emphasis throughout the guideline is a focus on multidisciplinary and co-ordinated assessment and management. The core MDT should consist of professionals with expertise in MND. This team should also have access to other specialties such as psychology, gastroenterology and a community team, when required. Supportive evidence for MDT working was found in cohort studies that compared MDT care with general neurology care. In each of these studies there was significant risk of bias and imprecision. The guidance suggested that MDT care increased survival by eight months. However, the use of riluzole and non-invasive ventilation (NIV) was also increased in this patient group.7

As part of the MDT, it is recommended that there is access to professionals with knowledge of palliative care in MND. End-of-life care is also a key focus of the guideline and opportunities for this to be discussed should be offered. That also includes discussion regarding advance care planning. This may be required early in patients with communication and cognitive difficulty.

For management of specific symptoms, it is stressed that consideration of any treatment or intervention should always be alongside a patient’s wishes and individual requirements. Many medications recommended in the guideline come from consensus recommendations extrapolated from their use in other neurological conditions. In spasticity, there was no evidence found for use of pharmacological treatments in MND and therefore the guidelines recommended the use of standard treatments and referral to specialist spasticity service if needed. For management of saliva, the recommendation of a first-line of anticholinergic comes from standard practice in other conditions such as Parkinson’s disease and cerebral palsy. The only trial performed in MND studied the use of botulinum toxin8 – this showed an improvement from a patient and carer perspective. However, this did not lead to improved quality of life or patient’s function. Botulinum toxin has been recommended as a second-line intervention as there is evidence of benefit in studies involving different neurological conditions.

As part of the ongoing assessment, the MDT should monitor for any changes in activities of daily living and equipment that would therefore be required. All equipment, such as home adaptations or wheelchair services, should be provided in a timely manner to maximise quality of life. Ability to use the equipment should also be reviewed regularly as the MND progresses. Although every patient has different needs, there is specific equipment that may be used more by patients with MND, such as mobile arm supports, collars and powered wheelchairs. Augmentative and alternative communication (AAC) aids are recommended when appropriate to an individual, bearing in mind that alternative low-level technology may be helpful.

The recommendations for use of NIV remain the same as in the 2010 guidance. The new addition to the guidance is on support available when NIV is to be stopped. This includes awareness of the legal and ethical responsibilities and knowledge of advance directives and lasting Power of Attorney.

Conclusions
The MND Association has welcomed this new guideline, especially the emphasis on MDT care, anticipatory assessments and recognition of cognitive changes. We also feel that these guidelines will be a very valuable tool for commissioners and service providers to ensure the provision of a coordinated service.

Clinicians should welcome the focus on symptomatic management and the use of data extrapolated from the use of interventions in other neurological conditions. Such a common-sense approach is particularly helpful in these times of austerity when, occasionally, effective symptomatic management is rejected by commissioners because of the lack of specific evidence of its efficacy in a particular illness. We also hope that increasing coordination between the neurology and palliative care services can provide a nucleus for further evolution of palliative neurology as an important sub-specialty with the potential to help many patients with diverse neurological conditions in the final stages of their illnesses, when the focus should shift to comfort and palliation.

Key points of new NICE MND guideline
• Importance of a co-ordinated MDT approach.
• Prompt recognition of possible diagnosis and early referral to specialist services.
• Consideration of cognitive impairment as both a presenting symptom and also as an important cause of disability.
• Recommendations for symptomatic treatments are mainly extrapolated from evidence related to other neurological conditions.
• Early consideration of palliative care, advance directive and end-of-life planning.
• Any intervention including equipment and/or communication aids should be supplied without delay.
• Care should be tailored around patient wishes.
• Consideration of family/carers at all points during MDT care.
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Declaration of interests
No conflicts of interest were declared.

References

Clinical question
Is cognitive behavioral therapy effective for adolescents with depression who decline antidepressant drug treatment?

Bottom line
In adolescents who eschew drug treatment of major depression, short-term cognitive behavioral therapy (CBT) is more effective than treatment as usual in inducing recovery, with a number needed to treat of 4 to 10. CBT also produced faster results. (LOE = 1b)

Reference

Study design: Randomized controlled trial (non-blinded) Funding source: Government Allocation: Uncertain Setting: Outpatient (primary care)

Synopsis
These researchers identified potential patients by mailing study brochures to parents of adolescents 12 to 18 years of age who had a recent prescription for an antidepressant (from a health maintenance organization in the United States) that went unfilled or was initially dispensed but not refilled. In other words, the patients did not fail antidepressant therapy but simply chose not to begin (or continue) it. The 212 adolescents who had major depressive disorder were randomized (allocation concealment uncertain) to continue treatment as usual (as determined by their primary care provider) or treatment as usual plus at least 4 sessions of CBT aimed at addressing unrealistic thinking and increasing pleasant activities (behavioral activation). The patients could continue a second set of 4 to 6 sessions, if desired, and most did. Recovery, defined as at least 8 weeks of well time as measured by the Children’s Depression Rating Scale—Revised, occurred significantly faster in the CBT group and was significantly more likely in the first year but not the second year of follow-up. Quality of life was better with therapy in the first year after therapy but not in the second year. Hospital admissions for psychiatric diagnoses were significantly higher in the control group. Substance use, suicidal behavior, and parent-reported outcomes were not different between the 2 groups, but the study may not have been long enough to find a difference if one exists. These results jibe with the results of several meta-analyses examining the effect of cognitive behavioral therapy for adolescents with depression.

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