

Quick reference guide

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Motor neurone disease

The use of non-invasive ventilation in the management
of motor neurone disease

About this booklet

This is a quick reference guide that summarises the recommendations NICE has made to the NHS in 'Motor neurone disease: the use of non-invasive ventilation in the management of motor neurone disease' (NICE clinical guideline 105).

Who should read this booklet?

This quick reference guide is for healthcare professionals and other staff who care for people with motor neurone disease.

Who wrote the guideline?

The guideline was developed by the Centre for Clinical Practice at NICE following the short clinical guideline process. The Centre worked with an independent group of healthcare professionals (including consultants, GPs and nurses), patients and carers, and technical staff, who reviewed the evidence and drafted the recommendations. The recommendations were finalised after public consultation.

For more information on how NICE clinical guidelines are developed, go to www.nice.org.uk

National Institute for Health and Clinical Excellence

MidCity Place
71 High Holborn
London
WC1V 6NA

www.nice.org.uk

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NICE clinical guidelines are recommendations about the treatment and care of people with specific diseases and conditions in the NHS in England and Wales.

This guidance represents the view of NICE, which was arrived at after careful consideration of the evidence available. Healthcare professionals are expected to take it fully into account when exercising their clinical judgement. However, the guidance does not override the individual responsibility of healthcare professionals to make decisions appropriate to the circumstances of the individual patient, in consultation with the patient and/or guardian or carer, and informed by the summary of product characteristics of any drugs they are considering.

Implementation of this guidance is the responsibility of local commissioners and/or providers. Commissioners and providers are reminded that it is their responsibility to implement the guidance, in their local context, in light of their duties to avoid unlawful discrimination and to have regard to promoting equality of opportunity. Nothing in this guidance should be interpreted in a way that would be inconsistent with compliance with those duties.

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Introduction

Motor neurone disease (MND) is a fatal neurodegenerative disease. It is characterised by the onset of symptoms and signs of degeneration of primarily the upper and lower motor neurones. Respiratory muscle weakness resulting in respiratory impairment is a major feature of MND, and is a strong predictor of quality of life and survival. Non-invasive ventilation can improve the symptoms and signs related to respiratory impairment and hence survival.

This guidance covers the identification and assessment of respiratory impairment in patients with MND, as well as provision of non-invasive ventilation. Differences in the pathway of care for patients with severe bulbar impairment or severe cognitive problems that may be related to respiratory impairment are included.

Patient-centred care

Treatment and care should take into account patients' individual needs and preferences. Good communication is essential, supported by evidence-based information, to allow patients to reach informed decisions about their care. Follow advice on seeking consent from the Department of Health or Welsh Assembly Government if needed. If the patient agrees, families and carers should have the opportunity to be involved in decisions about treatment and care.

Key to terms

FVC: forced vital capacity

MIP: maximal inspiratory pressure (P_Imax)

PaCO₂: arterial partial pressure of carbon dioxide

SNIP: sniff nasal inspiratory pressure

SpO₂: oxygen saturation measured by pulse oximetry

VC: vital capacity

Principles of care

Multidisciplinary team

- A multidisciplinary team should coordinate and provide ongoing management and treatment for patients with MND, including regular respiratory assessment and provision of non-invasive ventilation.
 - The team should be led by a healthcare professional with a specific interest in MND. The leader should ensure that the patient’s multidisciplinary care plan (see page 9) is coordinated and communicated to relevant healthcare and social care professionals, including the patient’s primary care team, as well as to the patient and (where appropriate) their family and carers.
 - The team should include a neurologist, a respiratory physician, an MND specialist nurse, a respiratory specialist nurse, a specialist respiratory physiotherapist, a respiratory physiologist, a specialist in palliative care and a speech and language therapist (team members do not have to be at the same location).
 - Access to other healthcare professionals should be provided as needed.
 - Team members who provide non-invasive ventilation should have appropriate competencies.

Information and support

- Offer to discuss the possible use of non-invasive ventilation with the patient and (if the patient agrees) their family and carers, at an appropriate time and in a sensitive manner. Provide information that is appropriate to the stage of the patient’s illness.

Offer discussions at one or more of the following times:

- soon after MND is first diagnosed
- when monitoring respiratory function (see pages 6–7)
- when respiratory function deteriorates
- if the patient asks for information.

Include information on:

- possible symptoms and signs of respiratory impairment (see table on page 6)
- the natural progression of MND and what to expect in the future
- the purpose, nature and timing of respiratory function tests, and explanations of the results
- available interventions for managing respiratory impairment, including the benefits and limitations of each
- accessing and using respiratory equipment, including that for non-invasive ventilation
- how non-invasive ventilation (as a treatment option) can improve symptoms associated with respiratory impairment and can be life prolonging, but does not stop progression of the underlying disease
- how non-invasive ventilation can be withdrawn
- palliative strategies as an alternative to non-invasive ventilation.

- Inform all relevant healthcare professionals about key decisions reached with the patient and their family and carers.
- Provide the patient and their family and carers with support and assistance to manage non-invasive ventilation. This should include:
 - training on using non-invasive ventilation and ventilator interfaces, for example:
 - ◆ emergency procedures
 - ◆ night-time assistance if the patient is unable to use the equipment independently (for example, emergency removal or replacement of interfaces)
 - ◆ how to use the equipment with a wheelchair or other mobility aids if required
 - ◆ what to do if the equipment fails
 - assistance with secretion management
 - information on general palliative strategies
 - an offer of ongoing emotional and psychological support¹ for the patient and their family and carers.
- Ensure that families and carers have:
 - an initial assessment if the patient they care for decides to use non-invasive ventilation, including:
 - ◆ their ability and willingness to assist in providing non-invasive ventilation
 - ◆ their training needs
 - the opportunity to discuss any concerns with members of the multidisciplinary team and/or other healthcare professionals.
- Discuss end-of-life care at an appropriate time – see page 10 for details.

¹ See 'Improving supportive and palliative care for adults with cancer'. NICE guidance on cancer services (2004). Available from www.nice.org.uk/guidance/csgsp

Identifying and assessing respiratory impairment

Symptoms and signs

Routinely monitor the symptoms and signs listed in the following table.

Symptoms	Signs
Breathlessness	Increased respiratory rate
Orthopnoea	Shallow breathing
Recurrent chest infections	Weak cough ²
Disturbed sleep	Weak sniff
Non-refreshing sleep	Abdominal paradox (inward movement of the abdomen during inspiration)
Nightmares	Use of accessory muscles of respiration
Daytime sleepiness	Reduced chest expansion on maximal inspiration
Poor concentration and/or memory	
Confusion	
Hallucinations	
Morning headaches	
Fatigue	
Poor appetite	

Respiratory function tests

A healthcare professional from the multidisciplinary team who has appropriate competencies should perform respiratory function tests at the following times (or arrange for them to be performed):

- As part of the initial assessment to diagnose MND, or soon after diagnosis (to establish baseline function).
- Every 3 months, but possibly more or less often depending on:
 - whether there are any symptoms and signs of respiratory impairment (see table above)
 - the rate of progression of MND
 - the patient's preference and circumstances.

Respiratory function tests and patients with a diagnosis of dementia

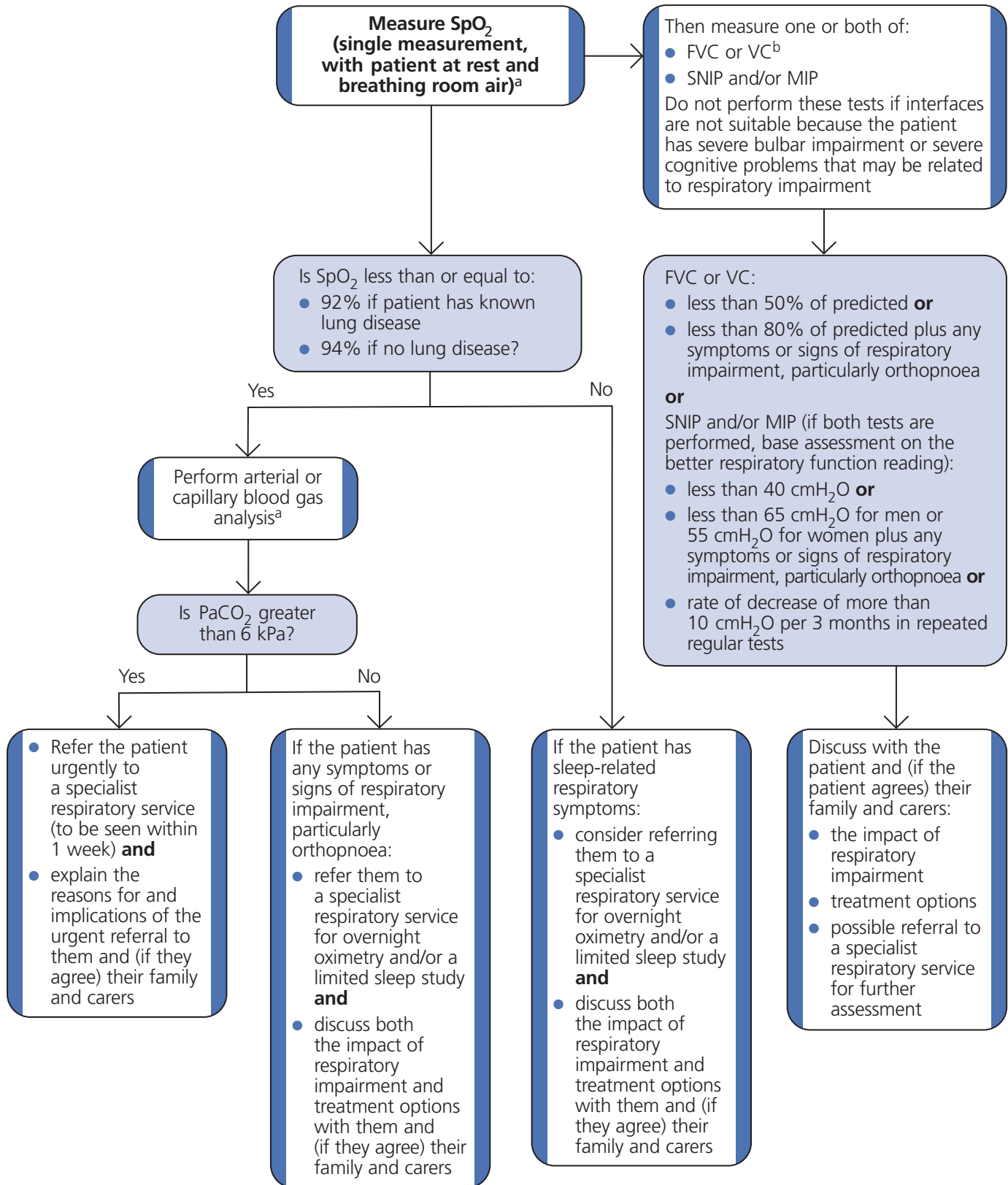
Base decisions on respiratory function tests on considerations specific to the patient's needs and circumstances, such as:

- their ability to give consent³
- their understanding of the tests
- their tolerance of the tests and willingness to undertake them
- the impact on their family and carers
- whether they are capable of receiving non-invasive ventilation.

² Weak cough could be assessed by measuring cough peak flow.

³ See 'Dementia'. NICE clinical guideline 42 (2006). Available from www.nice.org.uk/guidance/CG42

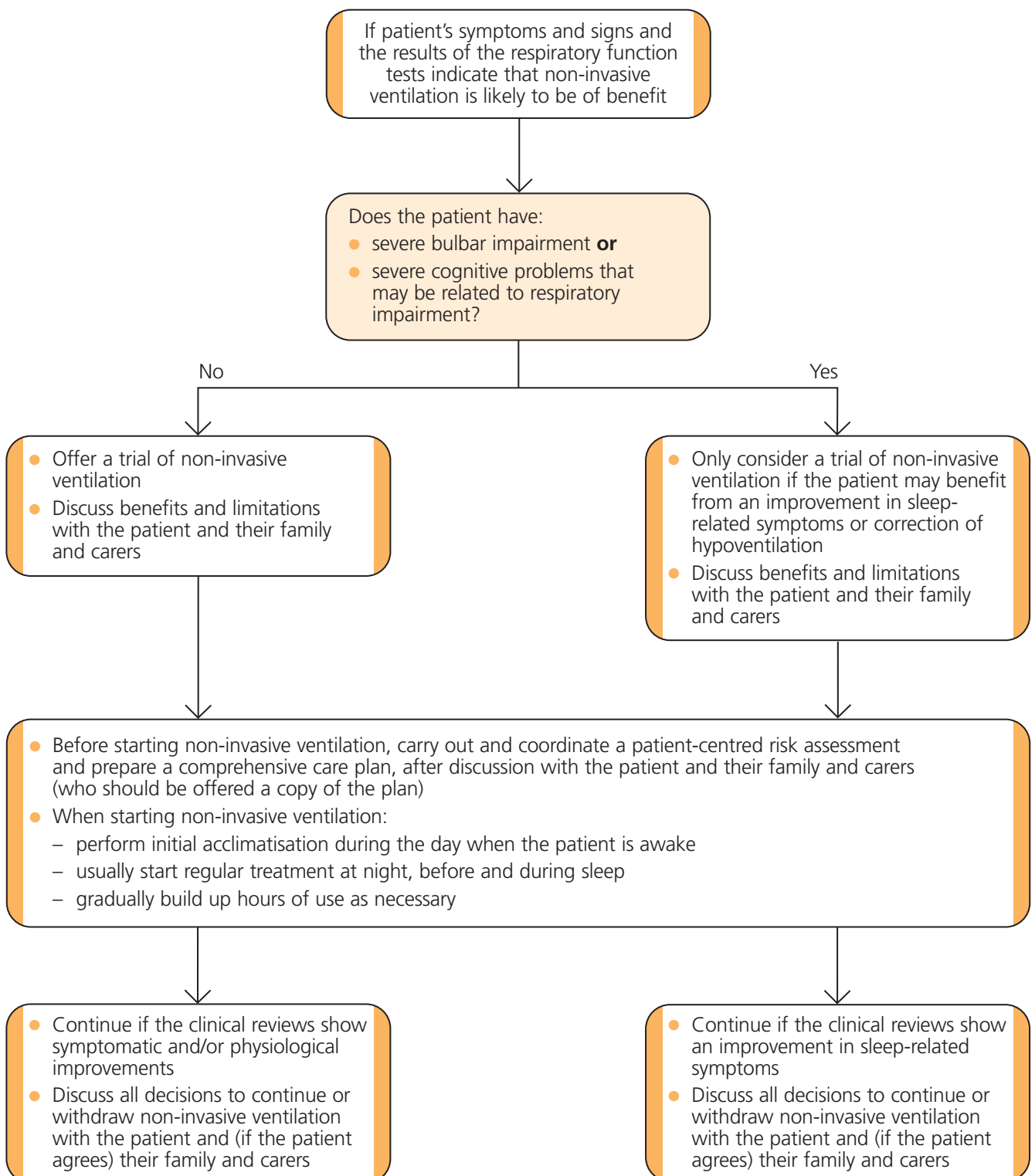
Assessment pathway



^a If it is not possible to perform pulse oximetry and/or arterial or capillary blood gas analysis locally, refer the patient to a specialist respiratory service.

^b The difference between the measurement of VC or FVC is very subtle and so either can be used.

Non-invasive ventilation



Risk assessment and care plan

The risk assessment should consider:	The comprehensive care plan should cover:
<ul style="list-style-type: none"> ● the most appropriate type of non-invasive ventilator and interfaces, based on the patient's needs and lifestyle factors ● the patient's tolerance of the treatment ● the risk, and possible consequences, of ventilator failure ● the power supply required, including battery back-up ● how easily the patient can get to hospital ● risks associated with travelling away from home (especially abroad) ● whether a humidifier is required ● issues relating to secretion management ● the availability of carers. 	<ul style="list-style-type: none"> ● long-term support provided by the multidisciplinary team ● the initial frequency of respiratory function tests and monitoring of respiratory impairment ● the frequency of clinical reviews of symptomatic and physiological changes ● the provision of carers ● arrangements for device maintenance and 24-hour emergency clinical and technical support ● secretion management and respiratory physiotherapy assessment, including cough-assist therapy (if required) ● training in and support for the use of non-invasive ventilation for the patient and their family and carers ● regular opportunities to discuss the patient's wishes in relation to continuing or withdrawing non-invasive ventilation, and other end-of-life considerations (see also page 10).

Non-invasive ventilation and patients with a diagnosis of dementia

Before a decision is made on the use of non-invasive ventilation, the neurologist from the multidisciplinary team should carry out an assessment that includes:

- the patient's capacity to make decisions and give consent⁴
- the severity of dementia and cognitive problems
- whether the patient is likely to accept treatment
- whether the patient is likely to achieve improvements in sleep-related symptoms and/or behavioural improvements
- a discussion with the patient's family and/or carers (with the patient's consent if they have the capacity to give it).

⁴ See 'Dementia'. NICE clinical guideline 42 (2006). Available from www.nice.org.uk/guidance/CG42

Planning end-of-life care

Offer to discuss end-of-life care with the patient and (if the patient agrees) their family and carers, at an appropriate time and in a sensitive manner.

Offer discussions at one or more of the following times:	Include information on:
<ul style="list-style-type: none"> ● around the time that MND is first diagnosed (but only if requested by the patient explicitly, or if the patient's clinical condition indicates that ventilator support will be needed in the immediate future) ● when non-invasive ventilation is accepted or declined ● when the patient is becoming increasingly dependent on non-invasive ventilation ● if the patient asks for information. 	<ul style="list-style-type: none"> ● planning end-of-life care ● considering advance decisions to refuse treatment ● considering what to do if non-invasive ventilation fails because of either: <ul style="list-style-type: none"> – an acute, but potentially reversible, deterioration in health or – irreversible disease progression ● strategies to withdraw non-invasive ventilation if the patient wishes ● the involvement of family and carers in decision making (with the patient's consent if they have the capacity to give it).

Further information

Ordering information

You can download the following documents from www.nice.org.uk/guidance/CG105

- The full guideline – all the recommendations, details of how they were developed, and reviews of the evidence they were based on.
- A quick reference guide (this document) – a summary of the recommendations for healthcare professionals.
- ‘Understanding NICE guidance’ – a summary for patients and carers.

For printed copies of the quick reference guide or ‘Understanding NICE guidance’, phone NICE publications on 0845 003 7783 or email publications@nice.org.uk and quote:

- N2228 (quick reference guide)
- N2229 (‘Understanding NICE guidance’).

Implementation tools

NICE has developed tools to help organisations implement this guidance (see www.nice.org.uk/guidance/CG105).

Related NICE guidance

For information about NICE guidance that has been issued or is in development, see www.nice.org.uk

Published

- Dementia. NICE clinical guideline 42 (2006). Available from www.nice.org.uk/guidance/CG42
- Improving supportive and palliative care for adults with cancer. NICE guidance on cancer services (2004). Available from www.nice.org.uk/guidance/csgsp

Updating the guideline

This guideline will be updated as needed, and information about the progress of any update will be available at

www.nice.org.uk/guidance/CG105

**National Institute for
Health and Clinical Excellence**

MidCity Place
71 High Holborn
London
WC1V 6NA

www.nice.org.uk

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