NICE guideline NG42
Motor neurone disease: assessment and management

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NICE & MND

NICE technology guidance (TA20) on the use of Riluzole for the treatment of motor neurone disease (published January 2001)

Published MND guidelines

EFNS guidelines on the clinical management of amyotrophic lateral sclerosis (MALS) – revised report of an EFNS task force (2012)

Riluzole dispensed in the community in primary care in England

Source: PCA

NICE Guidance
NIV in MND

Prior to 1999 only 28% of patients with dyspnoea and only 9.2% of those with FVC <40% were receiving NIV in the US (Miller 1999)

Only 5.5% of 2280 MND patients under review were receiving NIV in the UK in 2000 (Bourke 2002)

Now 28% of MND patients with respiratory impairment are said to receive NIV (2016 MND Care survey, 919 respondents)
Guideline scope

- Recognition and referral
- Information provided
- Prognostic factors
- Organisation of care
- Psychosocial support
- End of life care
- Symptom management
- Nutrition
- Communication
- Respiratory impairment and non-invasive ventilation
NICE MND Quality Standards

8 statements designed to drive measurable improvements in the 3 dimensions of quality:

- Patient safety
- Patient experience
- Clinical effectiveness
Guideline development

Guideline Development Group
- Multidisciplinary
- Patients and carers

National Clinical Guideline Centre technical team
- Information scientists, systemic reviewers
- Health economists
- Project management
- Editors

GRADE methodology of randomised controlled trials and cohort studies. In the absence of evidence recommendations were made by consensus

An original health economic model was developed to examine the cost effectiveness of MDT care using data from systematic reviews undertaken on the clinical and economic literature
Guideline Development Group

Chair
Dr David Oliver, Consultant in Palliative Medicine

Members
Dr Robert Angus, Respiratory Physician
Julie Brignall-Morley, Community Matron
Roch Maher, Patient/ carer member
Rachael Marsden, MND Nurse Specialist
Jennifer Rolfe, Occupational Therapist
Sandra Smith, Patient/ carer member
Dr Annette Edwards, Consultant in Palliative Medicine

Co-opted members
Sharon Abrahams, Neuropsychologist
Angelina Brooks, Dietitian
Karen James, Social Worker

Dr Steven Bloch, Speech and Language Therapist
Caroline Brown, Physiotherapist
Dr Chris McDermott, Consultant Neurologist
Dr Aleksandar Radunovic, Consultant Neurologist
Dr Ian Smith, Respiratory Physician
Dr Rachel Starer, General Practitioner
Jean Waters, Patient / carer member
Recognition and referral

- Protocol and pathway for referral
- Awareness of possible symptoms
- If suspected MND referral without delay
- Information for patient and family at all stages
2016 Improving MND Care survey (919 responses)

19% of people waiting a year or more to be referred to a Neurologist

Patients diagnosed in the last year significantly more likely to have been seen within a month of referral to a Neurologist than 3-10 years ago

- GPs giving better detail in referrals to Neurologists
- Improvement due to ? Red Flags tool

MND Care Centres & Networks significantly more likely to see people within a month of referral than non MND Association funded clinics
Painless, progressive weakness – Could this be Motor Neurone Disease?

1. Does the patient have one or more of these symptoms?

**Bulbar features**
- Dysarthria
- Slurred or quiet speech often when tired
- Dysphagia
- Liquids and/or solids
- Excessive saliva
- Choking sensation especially when lying flat
- Tongue fasciculations

**Limb features**
- Focal weakness
- Falls/trips – from foot drop
- Loss of dexterity
- Muscle wasting
- Muscle twitching/fasciculations
- Cramps
- No sensory features

**Respiratory features**
- Hard to explain respiratory symptoms
- Shortness of breath on exertion
- Excessive daytime sleepiness
- Fatigue
- Early morning headache
- Orthopnoea

**Cognitive features** (rare)
- Behavioural change
- Emotional lability (not related to dementia)
- Fronto-temporal dementia

2. Is there progression?

**Supporting factors**
- Asymmetrical features
- Age – MND can present at any age
- Positive family history of MND or other neurodegenerative disease

**Factors NOT supportive of MND diagnosis**
- Bladder/bowel involvement
- Prominent sensory symptoms
- Double vision/PTosis
- Improving symptoms

If yes to 1 and 2, query MND and refer to Neurology
If you think it might be MND please state explicitly in the referral letter. Common causes of delay are initial referral to ENT or Orthopaedic services.

Additional resources for GPs are available at www.mndassociation.org/gp
To order extra copies, please contact us by phone on 01604 611684 or email tina.downs@mndassociation.org

MND Red Flags

**Bulbar features**
- 25% of patients present with bulbar symptoms
  - Dysarthria
  - Quiet, hoarse or altered speech
  - Slurring of speech when often tired
  - Dysphagia – more often liquids first and later solids. Initially can be sensation of catching in throat or choking when drinking quickly
  - Excessive saliva
  - Choking sensation when lying flat
  - Weak cough – often not noticed by the patient
  - Painless progressive dysarthria – consider neurological referral rather than ENT

**Limb features**
- 70% of patients present with limb symptoms
  - Focal weakness – painless with preserved sensation
  - Distal weakness
  - Falls/trips – from foot drop
  - Loss of dexterity eg problems with zips or buttons
  - Muscle wasting – hands and shoulders. Typically asymmetrical
  - Muscle twitching/fasciculations
  - Cramps

**Respiratory features**
- Respiratory problems are often a late feature of MND and an unusual presenting feature. Patients present with features of neuromuscular respiratory failure
  - Shortness of breath on exertion
  - Excessive daytime sleepiness
  - Fatigue
  - Early morning headache. Patients often describe a “muzziness” in the morning being slow to get going or as if hung over
  - Un-refreshing sleep
  - Orthopnoea
  - Frequent unexplained chest infections
  - Weak cough and sniff
  - Nocturnal restlessness and/or sweating

**Cognitive features**
- Frank dementia at presentation is rare. Cognitive dysfunction is increasingly recognised, as evidenced by:
  - Behavioural change such as apathy or lack of motivation
  - Difficultly with complex tasks
  - Lack of concentration
  - Emotional lability (not related to dementia)

Ask specifically about a family history of these features.

Development group for this resource:
- Keele (S. Dray, P. Mansfield, L. Hanley) MND Association (J. Bedford, H. Fairfield)
- Neurology (P. Calleja, C. McDermott, K. Marrions, R. Orr, F. Radosavljevic, J. Weatherley, A. Wilks) Palliative Medicine (J. Baker)

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MND Association Care Centres and Networks

London: Kings College Hospital 1993
Newcastle Royal Victoria 1994
Birmingham Queen Elizabeth Hospital 1995
Nottingham Queen’s Medical Centre 1995
South Wales Care Network 1996
Liverpool Walton Centre 1998
Manchester Hope Hospital 2002
Oxford John Radcliffe Hospital 2002
Sheffield Royal Hallam Hospital 2002
London: National Hospital for Neurology and Neurosurgery 2003
Cambridge Addenbrookes 2004

Northern Ireland – Belfast 2004
Royal Preston Hospital 2004
Southampton General Hospital 2006
South West Peninsula Care Network 2008
Leeds Teaching Hospital 2008
London: Barts 2009
Middleborough James Cook University Hospital 2011
Bristol Southmead Hospital 2012
Brighton (due 2016)
Information and support at diagnosis

Diagnosis to be given by a Consultant Neurologist
  ◦ With knowledge and expertise in MND

Ensure
  ◦ People are asked about their wishes for information and involvement of family / carers
  ◦ Information on MND is provided as they wish
I would be very grateful for your urgent review of this 88 year-old lady who has recently moved to the area. She has suspected MND with rapid progression of her symptoms.

Seen elsewhere in September and December 2014 with muscle weakness and fatigue. Referred for nerve conduction studies but moved area before these were done.

‘We therefore have no formal diagnosis but her condition has continued to deteriorate. She is now immobile and hoist-dependent for her care needs.’

‘Her speech and swallow are normal and her mental functioning appears to be intact. She has very little movement of her upper arms and she is able to lift her legs against gravity. She is fully aware of her likely diagnosis and poor prognosis.’
MND Clinic: history

Fit and well up to summer 2014 – living alone, swimming once per week, gardening

Pins and needles in hands, fatigue, balance poor ‘couldn’t turn quickly’, arms and legs became progressively weaker

Moved to live with brother and sister-in-law, December 2014 difficulty climbing stairs

Moved to nursing home due to increased dependency March 2015; wheelchair, hoist transfer, unable to feed self, breathless on completing sentences
MND Clinic: examination

Wheelchair-bound; no neck, facial or tongue weakness, no fasciculations

Tone flaccid with profound weakness proximally and distally in upper limbs; shoulder abduction 2/5 B/L; elbow flex 4/5 on R, 2/5 on L; elbow ext -4/5 on R, 2/5 on L; wrist extension 2/5 on R, 1/5 on L

Increased tone in lower limbs with hip flex 2/5 on R, 1/5 on L; hip ext 4/5 B/L; knee flex -4/5 on R, 3/5 on L; knee ext 4/5 on R, 2/5 on L; ankle df 2/5 B/L, ankle pf 4/5 B/L

Reflexes brisk in upper and lower limbs, clonus at left ankle; extensor plantar on left, mute plantar response on right

Sensory examination: Pin-prick reduced throughout arms, legs and trunk to a high cervical level
MND Clinic: neurophysiology

Normal conduction velocities with marked denervation changes in bilateral FDI (++) spontaneous activity on EMG, no voluntary MUAPs)

Chronic neurogenic changes in all muscles sampled in arms, legs and paravertebral muscles (voluntary MUPs large amplitude, polyphasic, long duration)

Right glossus EMG – occasional polyphasic motor units during voluntary activity

Interpretation – EMG evidence of widespread motor axonopathy/neuronopathy affecting upper and lower limb muscles and thoracic region and suspicious in bulbar area; however partial denervation changes are only seen in hand muscles.
MND Clinic: MRI cervical spine
MRI cervical spine axial views and CT cervical spine
MND Clinic: Intervention and progress

Anterior cervical C3/4 decompression

Follow-up NCS/EMG: Marked improvement: voluntary activation - occasional polyphasic MUPs in FDI, L Tib ant and R gastrocnemius with full interference pattern in right FDI, and mildly reduced in Left FDI

Ward-based rehabilitation, mobilising with frame with supervision by time of transfer to the Rehabilitation Centre

Outpatient clinic review (6 months after surgery): mobilising independently, discharged
94 patient-caregiver pairs, 50 unpaired patients and 19 unpaired caregivers

Evaluation of the physician:
- Time spent discussing the diagnosis
- SPIKES protocol (setting, perception, invitation, knowledge, empathy, strategy)

56% patients found the physician who broke the news as average (30.7), below average (8.6) or poor (16.4)

48% of caregivers rated the physician as poor (14.4), below average (4.8) or average (28.8)

Effective communication and greater time spent discussing the diagnosis correlated with higher patient/caregiver satisfaction
The diagnosis appointment
(Improving MND Care Survey, 2016)

<table>
<thead>
<tr>
<th>Statement</th>
<th>% Yes, definitely</th>
<th>% Yes, to some extent</th>
<th>% No, but I didn't mind</th>
<th>% No, but I would have liked this</th>
</tr>
</thead>
<tbody>
<tr>
<td>a) I was told in a private place behind closed doors (n=876)</td>
<td>85%</td>
<td>57%</td>
<td>14%</td>
<td>3%</td>
</tr>
<tr>
<td>b) The diagnosis was explained in a way I could understand (n=851)</td>
<td>9%</td>
<td>31%</td>
<td>15%</td>
<td>14%</td>
</tr>
<tr>
<td>c) I was given the opportunity to ask questions (n=845)</td>
<td>71%</td>
<td>21%</td>
<td>4%</td>
<td>3%</td>
</tr>
<tr>
<td>d) I understood the answers to my questions (n=832)</td>
<td>57%</td>
<td>34%</td>
<td>6%</td>
<td>4%</td>
</tr>
<tr>
<td>e) The people giving the diagnosis understood how I might be feeling (n=843)</td>
<td>60%</td>
<td>26%</td>
<td>26%</td>
<td>12%</td>
</tr>
<tr>
<td>f) I felt supported throughout the consultation (n=868)</td>
<td>56%</td>
<td>19%</td>
<td>15%</td>
<td>14%</td>
</tr>
<tr>
<td>g) I was given time and space to think afterwards (n=849)</td>
<td>44%</td>
<td>16%</td>
<td>16%</td>
<td>14%</td>
</tr>
<tr>
<td>h) I was offered the opportunity for a follow up call or appointment with a neurologist soon after (n=837)</td>
<td>55%</td>
<td>14%</td>
<td>19%</td>
<td>14%</td>
</tr>
</tbody>
</table>
Prognostic factors

No clear prognostic factors

Shorter survival is associated with

- Bulbar presentation – speech and swallowing problems
- Weight loss
- Poor respiratory function
- Older age
- ALSFRS-R scale – lower scores
- Shorter time from first symptom to diagnosis
Organisation of care

Co-ordinated care from a clinic based MND multidisciplinary team

◦ Based in hospital or community
◦ Including health and social care professionals
  ◦ Expertise in MND
  ◦ Staff able to see people at home
◦ Ensuring communication
  ◦ All health and social care professionals / family / carers
◦ Co-ordinated assessments
  ◦ Every 2-3 months
  ◦ Ensuring co-ordinated care if person cannot attend clinic
Multidisciplinary team

Assessment, management and review of

- Weight, nutritional intake, feeding, swallowing
- Muscle problems – weakness, stiffness, cramps
- Physical function
- Saliva problems – drooling, thick saliva
- Speech and communication
- Cough effectiveness
- Respiratory function
Multidisciplinary team – areas to review

Pain and other symptoms
Cognition and behaviour
Psychological needs
Social care needs
End of life issues
Information and support needs
  ◦ Person with MND
  ◦ Family / carers
Multidisciplinary team – suggested skills

Neurologist
Specialist MND nurse
Dietitian
Physiotherapist
Occupational therapist
Respiratory healthcare professional
Speech and language therapist
Palliative care expertise – may be one of the team members
MDT access to other services

Established relationships with
- Clinical psychology / neuropsychology
- Social care
- Counselling
- Respiratory ventilation services
- Nutrition team (Gastroenterology and Interventional Radiology)
- Orthotics
- Wheelchair services
- Assistive technology services
- Alternative and augmentative technology services
- Community neurology teams
- Specialist palliative care
Multidisciplinary team

Regular assessments according to the person’s needs

See earlier if there are changes in condition
  • Responding to triggers for earlier assessment

Ensure all are informed of key decisions

As condition will deteriorate professionals not to discharge form caseloads to ensure continuity of care

Consider referral to specialist palliative care if complex needs
# MND care costs

<table>
<thead>
<tr>
<th>Disease state, cost per year (2014 GBP)</th>
<th>Mild</th>
<th>Moderate</th>
<th>Severe</th>
<th>Terminal</th>
</tr>
</thead>
<tbody>
<tr>
<td>Italy (2003)</td>
<td>£799</td>
<td>£2,196</td>
<td>£3,494</td>
<td>£5,041- £5,291</td>
</tr>
<tr>
<td>Holland (2003)</td>
<td></td>
<td></td>
<td></td>
<td>£4,685</td>
</tr>
<tr>
<td>Denmark (2009)</td>
<td></td>
<td></td>
<td></td>
<td>£9,456</td>
</tr>
<tr>
<td>Spain (2003)</td>
<td></td>
<td>£2,433</td>
<td></td>
<td>£3,980</td>
</tr>
<tr>
<td>UK (1996)</td>
<td>£1,978</td>
<td>£1,212</td>
<td>£2,671</td>
<td>£5,160</td>
</tr>
</tbody>
</table>
MDT care costs

MDT clinic:
- £101.01 per patient spent outside of patient contact, dedicated to MDT per 9 weeks annually
- £634.59 per patient spent at dedicated MDT patient meetings per 9 weeks annually

MDT extended outreach team:
- £540 per patient annually (6 visits per year, 3 hours spent in-between clinic visits)

Total per patient annually: £1,275.61 (range £547 - £2,888)

General Neurology care (2 visits per year: £352)
MDT care costs

Incremental cost-effectiveness ratio (ICER) for MDT, Riluzole and NIV use: £26,672 per QALY

ICER for the above but using patient-elicited VAS quality of life scores: £20,791 per QALY

ICER for the above but using patient-elicited standard gamble quality of life scores: £17,387 per QALY

ICER for the above with zero MDT costs: £19,045 per QALY
Information and support at diagnosis

Single point of contact for the MND MDT

Follow up appointment with a MDT member within 4 weeks

Refer to Social Services
  ◦ If there are social care needs
  ◦ Ensuring carers are aware of Carer’s Assessment

81% of the respondents had a named person responsible for coordinating their care and support (2016 Improving MND Care survey)
Cognitive assessment

People with MND and Frontotemporal dementia may lack capacity
- Mental Capacity Act
- http://www.gmc-uk.org/Mental_Capacity_flowchart

At diagnosis check for cognitive / behavioural changes
- Explore with person and family
- Refer for assessment if necessary in line with NICE guideline on dementia (CG42)

Tailor discussions to the person
- Communication ability
- Cognition
- Mental capacity
Psychological and social support

MDT assessment to include discussion of psychological / emotional impact of MND

Offer information on support
  ◦ Person with MND
  ◦ Family / carer

Social care to be discussed with experienced social care worker
  ◦ Personal care - provided with continuity of carers
  ◦ Finances
  ◦ Social activities / hobbies / social media
  ◦ Respite care
Planning for end of life care

Discussion on end of life care whenever the person asks
Support and advice on advance care planning
  ◦ What may happen
  ◦ Advance care plans
    • ADRT (Advance Directive to Refuse Treatment)
    • LPA (Lasting Power of Attorney)
  ◦ Anticipatory medication at home
  ◦ Specialist palliative care involvement
  ◦ The person’s wishes
    • Place of care
    • Place of death
    • What happens if deterioration / other illness
Planning for end of life care

As end of life approaches
- Provide additional support so family are bale to reduce responsibility and spend time with the person
- Ensure prompt access to
  - Communication aid to allow communication
  - Specialist palliative care
  - Equipment - syringe driver, beds, commodes, hoist
  - Anticipatory medication – “Just in case “

Bereavement care for families / carers
Muscle problems

Cramps
- First line
  - Quinine
- Second line
  - Baclofen
  - Tizanidine
  - Dantrolene
  - Gabapentin

Ensure medication is appropriate – eg swallowing issues
Review regularly for effectiveness / side effects
Muscle problems – exercise programmes

Aims
- Maintain joint range of movement
- Prevent contractures
- Reduce stiffness and discomfort

Can be resistance / active-assisted / passive

Check to see if family / carers can assist in the programme

May need referral for orthotics

Aims
- Maintain joint range of movement
- Prevent contractures
- Reduce stiffness and discomfort
Equipment provision

Physiotherapy and occupational therapy assessment

Consider
- Activities of daily living
- Mobility and prevention of falls
- Home environment and adaptation
- Assistive technology – environmental control

Provide equipment without delay to allow maximise daily living and independence

Equipment can meet changes as deterioration occurs

Ensure referral for wheelchair services and orthotics without delay

Ensure integration with other aids – eg AAC devices

Regular review
Communication

Assess needs for communication
- Face to face
- Remote – telephone/ email / social media

Provide equipment to allow person to participate in activities
- Low level - alphabet board, picture board
- High level – PC / Tablet based

Refer to Specialised NHS AAC Hub if complex equipment anticipated

Ensure equipment is integrated with all equipment provision
Symptom management - Saliva

Advice on posture / diet / swallowing / oral care

Watery saliva
- Antimuscarinic medication trial
  - Glycopyrrolate
  - Hyoscine hydrobromide
- Injection of Botulinum toxin into salivary glands
- Radiotherapy suitable only in rare cases

Thick saliva
- Stop medication that may thicken
- Advice on diet / posture
- Humidification / nebulisers / carbocisteine
Nutrition

From diagnosis assess weight, nutrition and swallowing

Assess ability to eat and drink
- Aids to help plate to mouth
- Food drink preparation
- Advice on positioning / seating / posture
- Coping with social situations

If there are suspected swallowing problems ensure a swallowing assessment
Nutrition

Assess swallowing
- Positioning / seating
- Modification of consistency
- Respiratory problems
- Risk of aspiration / choking
- Fear of choking
- Psychological issues – social situations

Discuss gastrostomy early and regularly
Nutrition

Explain benefits of early placement of gastrostomy
  ◦ Discussion of risks / benefits

Gastrostomy placement without delay when decision is made

If FTD
  ◦ Support of carers and explanation of increased risks
  ◦ Gastrostomy may be considered but assessment of the person being able to accept / cope with the placement and the gastrostomy must be made
Respiratory impairment in MND

<table>
<thead>
<tr>
<th>SYMPTOMS</th>
<th>SIGNS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Breathlessness</td>
<td>Increased respiratory rate</td>
</tr>
<tr>
<td>Orthopnoea</td>
<td>Shallow breathing</td>
</tr>
<tr>
<td>Recurrent chest infections</td>
<td>Weak cough*</td>
</tr>
<tr>
<td>Disturbed sleep</td>
<td>Weak sniff</td>
</tr>
<tr>
<td>Non-refreshing sleep</td>
<td>Abdominal paradox (inward movement of the abdomen during inspiration)</td>
</tr>
<tr>
<td>Nightmares</td>
<td>Use of accessory muscles of respiration</td>
</tr>
<tr>
<td>Daytime sleepiness</td>
<td>Reduced chest expansion on maximal inspiration</td>
</tr>
<tr>
<td>Poor concentration and/or memory</td>
<td></td>
</tr>
<tr>
<td>Confusion</td>
<td></td>
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<tr>
<td>Hallucinations</td>
<td></td>
</tr>
<tr>
<td>Morning headaches</td>
<td></td>
</tr>
<tr>
<td>Fatigue</td>
<td></td>
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<tr>
<td>Poor appetite</td>
<td></td>
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</tbody>
</table>

* Weak cough could be assessed by measuring cough peak flow.
Reduced respiratory function - testing

Pulse oximetry - SpO2
  ◦ Overnight oximetry

Forced vital capacity (sitting and supine)
Sniff Nasal Inspiratory pressure – SNIP
Arterial / capillary blood analysis
Cough peak flow
Respiratory function

Assess regularly
- Exclude reversible causes – eg infection

Offer non-invasive ventilation if there is respiratory impairment

Urgent use of NIV if worsening respiratory failure

Opioids / benzodiazepines may be used to relieve breathlessness
Cough effectiveness

Cough augmentation techniques should be offered if the person cannot cough effectively

- Breath stacking and/or manual assisted cough
- If bulbar dysfunction breath stacking ineffective
  - Assisted breath stacking – using lung volume recruitment bag
- Mechanical cough assist considered
  - Breath stacking ineffective and/or
  - During respiratory infection
Non-invasive ventilation

Discuss regularly at appropriate times, sensitively
- Soon after diagnosis of MND
- When monitoring respiratory function
- When respiratory function deteriorates
- If person asks

Discussion should provide information on
- Possible signs and symptoms of respiratory impairment
- Role of monitoring and explanation of results
- The use of NIV to relieve symptoms and may prolong life, but does not stop the underlying progression of MND
Non-invasive ventilation

Discussion of management of breathlessness
- NIV has advantages and disadvantages
- Dependency on NIV is possible
- Options for treating infections
- Support of how to cope if there is a distressing situation and deterioration
- Effectiveness of medication in helping breathlessness – opioids
- Psychological techniques and support
NIV – check person does understand

- What NIV is and what it can achieve
- Other options for breathlessness – opioids
- The issues of compatibility with other equipment – eg eye gaze
- The need for carers to be able to help with NIV and have some training in ensuring it works effectively
- NIV can be stopped at their request at any time
  - If they become dependent on it
  - Medication may be needed to relieve symptoms as they may become distressed if it is stopped
Respiratory symptoms – use of NIV

Offer a trial of non-invasive ventilation
◦ If there is likely to be a benefit for the person

Consider trial of NIV in people with severe bulbar problems or severe cognitive problems
◦ If it is thought they would cope with NIV and this may help in sleep-related problems or hypoventilation
NIV – issues to consider

Before starting NIV risk assessment by the MDT

◦ Most appropriate ventilator and interface
◦ Tolerance to NIV
◦ Risk of ventilator failure
◦ Power supply needed - including need for battery back up
◦ How easy for the person to reach a hospital for help
◦ Risk of travelling abroad if they wish to do so
◦ Need for humidification
◦ Assessment of secretions / saliva management
◦ Availability of help from carers
NIV – issues to consider

Before staring NIV ensure there is a plan for

- Support from MDT
- Frequency of testing and monitoring
- Clinical reviews
- Carer support
- Technical support of the ventilator and equipment
- Training for person and family
- Discussion of potential dependency and wishes of person to continuation or withdrawal of NIV
When to start NIV

Symptoms related to respiratory muscle weakness

Evidence of significant respiratory weakness: FVC less than 80%, supine VC less than 25% of sitting or standing VC, SNP < 40cms H₂O

Evidence of either morning hypercapnia (PCO₂ greater of 6.5kPa) or significant nocturnal desaturation (less than 90% for > 5% of sleep time)
Starting NIV

Initial acclimatisation in the day
Start regular use at night
Increase slowly in use

Provide training for person and family
- Emergency situations
- Use in a wheelchair
- Secretion management
- Palliative care strategies – eg use of opioids
NIV and FTD

Careful MDT assessment
- Person’s capacity to decide themselves
- Acceptability of NIV to person
- Is NIV likely to improve symptoms
- Wider discussion with family / carers
- Consider medication to help symptoms
  - Opioids
  - Benzodiazepines
Stopping NIV

Careful consideration of the plan to stop – considering practical, ethical and legal aspects

Ensure there is support

- From professionals who have expertise in
  - Stopping ventilation
  - Palliative medication use
  - Supporting the person, family/carers / health and social care professionals
  - Legal and ethical aspects

### NIV costs per 100,000 population

<table>
<thead>
<tr>
<th>Recommendations with significant costs</th>
<th>Costs (£ per year)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Year 1</td>
</tr>
<tr>
<td>Initiation of NIV</td>
<td>1000</td>
</tr>
<tr>
<td>Equipment for NIV</td>
<td></td>
</tr>
<tr>
<td>– additional single ventilators</td>
<td>6000</td>
</tr>
<tr>
<td>Equipment for NIV</td>
<td></td>
</tr>
<tr>
<td>– additional second ventilators</td>
<td>0</td>
</tr>
<tr>
<td>Estimated cost of implementation</td>
<td>7000</td>
</tr>
</tbody>
</table>
NICE NG42: Conclusions

A comprehensive clinic based multidisciplinary approach is effective in supporting patients and families and may extend life as well as improve quality of life. Ongoing assessment of all areas of care is essential throughout the disease progression.
Recommendations for research

Organisation of care
Cognitive assessment
Prognostic tools
Management of sialorrhoea
Nutrition
Augmentative and alternative communication
NICE NG42

Further information: www.nice.org.uk/NG42