Respiratory weakness can develop at any stage of disease progression and may cause shortness of breath, fatigue, impaired quality of life and somnolence. Dyspnoea is caused by weakened respiratory muscles – intercostals, diaphragm and abdominal muscles.

The diagnosis and management of respiratory insufficiency is critical because most deaths from ALS are due to respiratory failure (Miller and others 2009a).

Rafiq and others 2012

Respiratory muscle function significantly predicts survival and quality of life in patients with MND. The management of respiratory symptoms and maintenance of lung compliance are particularly important. NIV prolongs life and maintains quality of life in patients with MND. NIV requires a specialist service with ability to monitor and detect hypoventilation, to start NIV with the optimal machine and interface and optimised settings, to monitor the effectiveness of NIV and to manage complications. It is essential to have effective communication with the patient and family throughout the service. It is particularly important that the patient understands the role of NIV, and that the clinician records their wishes early on regarding management of intercurrent illness, increasing reliance of NIV and end-of-life care. The recent NICE guidelines concluded that such a service is a cost-effective use of NHS resources.

Practice points

- Domiciliary NIV therapy has been an important advance for patients with MND.
- Cough strength may be impaired in MND; a suitable cough augmentation technique can help such patients.
- Clinicians should pay careful attention to nutritional support in patients with MND.
- Pharmacological management of symptoms may include anticholinergics for excess saliva and mucolytics, for example, carbocisteine, to reduce secretion viscosity.
- Tracheostomy ventilation is an option for patients who wish to live but who cannot use NIV.
- Palliative care needs, end-of-life issues and stopping respiratory support should be discussed regularly with the patient and the family.

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Clinical features

- early in the course of the disease nocturnal hypoventilation may not be manifest by obvious shortness of breath but more by headaches or general tiredness
- later, more marked shortness of breath may appear but this tends to be at the time when the person living with MND has severe generalised weakness which may affect the throat and limb muscles - the symptoms at that time may include:
  - breathlessness on exertion, sometimes just the exertion of speaking or eating
  - sleep disturbance, anxiety and panic
  - orthopnoea - breathless lying flat
- quiet voice and fewer words per breath
- weakened cough and sneeze
- hypoventilation is worse during sleep. Upper airway may also be partially obstructed due to bulbar and laryngeal muscle weakness
- increasing blood CO2 levels result in headaches, nausea and tiredness, especially on waking
- hypoxia, especially if there is coexisting lung disease

Management

Regular respiratory assessments, including sleep studies, by a respiratory physician are indicated in most persons living with MND.

- these should be initiated at a relatively early stage and their timing depends upon the level of abnormality detected
- where nocturnal hyperventilation is detected it may be appropriate for the respiratory physician to initiate and prescribe non-invasive positive pressure ventilation (NIPPV)

Respiratory management algorithm

(Miller and others 2009a)

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KEY

PFT = pulmonary function tests
PCEF = peak cough expiratory flow
NIV = noninvasive ventilation
SNP = sniff nasal pressure
MIP = maximal inspiratory pressure
FVC = forced vital capacity (supine or erect)
Abnl.nocturnal oximetry = pO2

*Symptoms suggestive of nocturnal hypoventilation: frequent arousals, morning headaches, excessive daytime sleepiness, vivid dreams.
+ If NIV is not tolerated or accepted in the setting of advancing respiratory compromise, consider invasive ventilation or referral to hospice.