Respiratory implications of motor neurone disease

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Spontaneous breathing overnight - air
Think of the diagnosis

"Pull out! Pull out! . . . You've hit an artery!"
Respiratory muscle involvement in MND

• 31/36 patients with MND had respiratory muscle weakness at presentation
• Only 7 complained of respiratory symptoms
• Rate of decline (tended to be linear but with great deal of inter patient variability)
  – VC -3.5% / month,
  – PImax +2.9 cm H₂O /month
  – PEmax -3.4 cm H₂O /month

Schiffman & Belsh Chest 1993; 103:508-13
<table>
<thead>
<tr>
<th>Breathlessness</th>
<th>Increased respiratory rate</th>
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<tr>
<td>Orthopnoea</td>
<td>Shallow breathing</td>
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<tr>
<td>Recurrent chest infections</td>
<td>Weak cough[^1]</td>
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<tr>
<td>Disturbed sleep</td>
<td>Weak sniff</td>
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<td>Non-refreshing sleep</td>
<td>Abdominal paradox (inward movement of the abdomen during inspiration)</td>
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<td>Nightmares</td>
<td>Use of accessory muscles of respiration</td>
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<td>Daytime sleepiness</td>
<td>Reduced chest expansion on maximal inspiration</td>
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<td>Poor concentration and/or memory</td>
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<tr>
<td>Confusion</td>
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<td>Hallucinations</td>
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<td>Morning headaches</td>
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<tr>
<td>Fatigue</td>
<td></td>
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<td>Poor appetite</td>
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Effects of non-invasive ventilation on survival and quality of life in patients with amyotrophic lateral sclerosis: a randomised controlled trial

Stephen C Bourle, Mark Tomlinson, Tim L Williams, Robert E Bullock, Pamela J Shaw, G John Gibson

Lancet Neurol 2006; 5: 140–47
Assisted ventilation in MND - problems

- life of marginal quality
- loss of dignity
- inability to communicate
- disruption to the lives of other family members
- difficulty in stopping high technology life sustaining care once it has been started
- for most it is not just a question of longer life, but better quality of life.
Duchenne MD (n=80)  Post polio (n=380)
Quality of life

• very difficult to measure
• highly individual
• horizons may change with time
• need to consider the main carer
Figure 3: Time SAQLI symptoms domain maintained above 75% of presandomisation assessment
A: all patients; B: patients with normal or moderately impaired bulbar function; C: patients with severe bulbar impairment. QoL=quality of life.
Motor neurone disease

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

Issued: July 2010

NICE clinical guideline 105
guidance.nice.org.uk/cg105
Multidisciplinary team

A multidisciplinary team should coordinate and provide ongoing management and treatment for a patient with MND, including regular respiratory assessment and provision of non-invasive ventilation.
Referral to Respiratory Service

- SpO2 < 92% if known chronic lung disease
- SpO2 < 94%
- Sleep related symptoms
- PaCO2 > 6 kPa
- PaCO2 < 6 kPa and symptoms – overnight oximetry or respiratory variable sleep study
Respiratory function tests: 1

- Assess baseline respiratory function at, or soon after, diagnosis
- Measure:
  - $\text{SpO}_2$ (measured by pulse oximetry)
  - then one or both of:
    - forced vital capacity or vital capacity
    - MIP and/or SNIP
- If the patient has severe bulbar impairment or severe cognitive problems, measure $\text{SpO}_2$ but omit other tests if interfaces are unsuitable
Respiratory function tests: 2

– Perform tests every 3 months, but this can vary depending on symptoms and signs, patient preference and rate of progression of MND

– Perform arterial or capillary blood gas analysis if SpO$_2$ is low

– Refer urgently to a specialist respiratory service if PaCO$_2$ is greater than 6 kPa
| Forced vital capacity (FVC) or vital capacity (VC) | Sniff nasal inspiratory pressure (SNIP) and/or maximal inspiratory pressure (MIP)  
(if both tests are performed, base the assessment on the better respiratory function reading) |
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<tr>
<td>• FVC or VC less than 50% of predicted value</td>
<td>• SNIP or MIP less than 40 cmH₂O</td>
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<tr>
<td>• FVC or VC less than 80% of predicted value plus any symptoms or signs of respiratory impairment (see recommendation 1.1.7), particularly orthopnoea</td>
<td>• SNIP or MIP less than 65 cmH₂O for men or 55 cmH₂O for women plus any symptoms or signs of respiratory impairment (see recommendation 1.1.7), particularly orthopnoea</td>
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<td>• Repeated regular tests show a rate of decrease of SNIP or MIP of more than 10 cmH₂O per 3 months</td>
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Secretion management

• Avoiding precipitants
  – Recognise and prevent aspiration
  – Hypersalivation
    • Suction
    • Atropine
    • Hyoscine
    • Glycopyrrholate
    • Botox
    • DXT
    • Tracheostomy

• Early antibiotics
Measuring Cough Peak Flow (PCF)

- Cheap
- Portable
- Simple to do
- Best of 3
Neuromuscular

Chatwin et al ERJ 2003
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

– Discuss:
  – planning
  – advance decisions to refuse treatment
  – what to do if non-invasive ventilation fails
  – strategies to withdraw non-invasive ventilation if the patient wishes
  – involvement of family and carers
Decision making in MND

• Patient
  – what are their goals
  – how far are they prepared to go
  – shifting horizons

• Carer
  – increasing burden
    • Ventilator
    • other care needs
ALS Association Data on 75 ALS patients using MV

- 25 used nIPPV - all started electively.
- 50 used tracheostomy-IPPV usually as a result of emergency hospitalization
- Survival with nIPPV ranged from 6 to 64 months
- 15 used nIPPV from 20 to 24 h daily
- 100% of the users were glad they chose nIPPV.
- 25 tIPPV patients (50%) lived in a sub-acute skilled nursing facility and only 18 of these (72%) were satisfied with their quality of life.
• 90% of patients were glad that they had chosen HMV and would do so again.
• 94% of carers were glad that the patient had chosen HMV and 83% would encourage the patient to choose it again.
• 50% said that they would choose HMV for themselves, citing not wanting to be a burden to others as the main reason.

Stopping treatment

• Seldom a problem with NIV
  – requires effort from patient to keep going
  – masterly inactivity easier than active withdrawal

• Tracheostomy
  – more difficult
  – our job is to prolong life not the process of dying
Welcome

Welcome to the Plattitude. If you are a regular visitor then click on the blog tab to read our latest news. If you are new to this site than here is a bit of background information about us and how we got to where we are now.

Thanks for taking the time to read about our experiences, your support and comments really do help to keep us going.

Neil, Louise & Oscar Platt.

A bit of background

Neil Platt and Louise Noble became Mr & Mrs Platt in November 2004 narrowly missing the Tsunami on their honeymoon in Sri Lanka.

After living and working in London for the next couple of years, the couple bought their first home together soon before the arrival of Oscar in August 2007.

Towards the end of 2007 Neil noticed that he was losing the power in one toe on his right foot. By Christmas he had started to use a stick to help him whilst walking. In the months following he went through a series of tests to discount any other cause of his symptoms and was finally diagnosed with Motor Neurone Disease in February of 2008.

Neil and Louise started writing this blog in July 2009 to keep all of their friends and family informed about how they were getting on and also in the knowledge that documenting the progress of Neil’s condition as openly as possible would help to promote an awareness of MND and could also help other sufferers of this awful disease.

Neil and Louise are now on a mission to raise as much public awareness of
‘Have a safe journey. I will see you some time’

The last moments: How Craigavon killed himself!

‘If I opt for life, then that is choosing to be tortured, rather than end this journey’

A Golf that’s economical even before you turn the ignition.

£750 to £1,400 off the Golf range.

Volkswagen

‘Her husband was with her the whole time until the final moment.’

‘I don’t think anyone would have predicted that,’ one friend said.
Conclusions

• Elective decision making better than crisis management.
• “Respiratory” symptoms not a good guide
• Serial measurements useful prognostically
  – pulmonary function tests
  – respiratory muscle tests
  – arterial blood gases
  – sleep monitoring
  – cough peak flow
Find out more

• Visit [www.nice.org.uk/CG105](http://www.nice.org.uk/CG105) for:
  – the guideline
  – the quick reference guide
  – ‘Understanding NICE guidance’
  – costing report and template
  – audit support