Chronic Non-invasive Ventilation for Neuromuscular Disorders: Evidence & Ethics

Dr Karen Detering
Sleep

- normal sleep is important to allow:
  - good health
  - normal cognitive function
  - normal quality of life

- sleep stages - REM and NREM
  - NREM (inactive mind, active body)
  - REM (active mind, inactive body)
Normal sleep
Control of Ventilation

- respiratory centre does not have a pacemaker
- during wakefulness ventilation is controlled by
  - respiratory centre input (brainstem)
    - chemical (central & peripheral chemoreceptors)
    - mechanical information (pulmonary & chest wall receptors)
  - higher cortical input (wakefulness drive)
    - important to allow e.g. speech, singing
Normal Sleep Neurophysiology

• during normal sleep:
  • loss of “wakefulness drive”
  • ↓ response to O2 & CO2 change - REM > NREM
  • change in muscle function - upper airway, chest wall and diaphragm
    • more severe in REM sleep
  • change in lung volumes - sleep and posture
Clinical Sequelae of Sleep

- ↓ ventilatory reserve & ↓ arousal responses
  - if pre existing lung disease or respiratory compromise
    - significant nocturnal hypoxaemia and hypercapnia
  - sleep - critical link in development of hypercapnic respiratory failure
  - as patients develop respiratory failure ⇒ deterioration which occurs - worsened by changes during sleep
Evolution of Respiratory Failure

- ↓ Ventilation ⇒ ↑CO₂ & ↓O₂
  - ↑ Arousals ⇒ sleep fragmentation
    - Depression of arousal response
      - Worsening hypoventilation
        - ↓ Respiratory drive
          - Nocturnal hypoventilation
            - daytime respiratory failure
## Important Symptoms

### GENERAL
- daytime fatigue
- reduced motivation
- breathlessness
- generally feeling unwell
- weight loss
- ↓ appetite
- ↓ exercise tolerance

### SLEEP FRAGMENTATION
- unrefreshing sleep
- daytime sleepiness
- ↓ cognitive function
- impaired alertness

### BREATHING DURING SLEEP
- morning headache
- progressive breathlessness
- orthopnoea
- wake choking or SOB @ night
- witnessed apnoeas
- snoring
# Conditions Where Ventilation May Be Indicated

<table>
<thead>
<tr>
<th>Neuromuscular Disorders</th>
<th>Disorders of ventilatory control</th>
<th>Lung disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Motor Neurone Disease</td>
<td>• obesity / hypoventilation</td>
<td>• COPD</td>
</tr>
<tr>
<td>• Muscular Dystrophy</td>
<td>• OSA with hypoventilation</td>
<td>• Cystic Fibrosis</td>
</tr>
<tr>
<td>• post polio syndrome</td>
<td>• brainstem injury</td>
<td>• bronchiectasis</td>
</tr>
<tr>
<td>• spinal cord injury</td>
<td>• primary alveolar hypoventilation</td>
<td></td>
</tr>
<tr>
<td>• other - diaphragm paralysis, spinal muscular atrophy</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

## Chest Wall Disorders
- kyphoscoliosis
- thoracoplasty
- pulmonary resection
Ventilation in Chronic Respiratory Failure

• symptoms & ↓ ventilation during sleep may be reversible
• nocturnal ventilation - support at most vulnerable time
  • associated with improvement in
    • ABGs
    • sleep quality
    • symptoms & daytime functioning
• some patients - use during day time as well
Treatment of Respiratory Failure with Ventilation

Assisting ventilation

↑ Ventilation ⇒ ↓ CO₂ & ↑ O₂

↓ Arousals ⇒ ↓ sleep fragmentation

Improvement of arousal response

Improved hypoventilation

↑ Respiratory drive

↓ Nocturnal hypoventilation

Improvement in daytime respiratory failure
Motor neurone disease – pre & post NPPV
Ventilation in Chronic Respiratory Failure

• in “at risk” patients look for;
  • symptoms of nocturnal hypoventilation
  • symptoms related to sleep fragmentation
• but symptoms can be non specific & often develop insidiously ⇒ need close follow up in at risk groups

• monitoring - to include assessment of blood gases, pulmonary function tests & ideally polysomnography
Goals of Care

• improving gas exchange & sleep quality
• control of symptoms
• improving function & quality of life
• reducing morbidity & reducing mortality
• other issues
  • staying in control
  • “good” death
  • support for family and loved ones
Ventilation in Neuromuscular disease

- difficulties
  - when to start treatment
  - when to stop treatment
  - should invasive ventilation be used?
- need to consider:
  - quality of life
  - difficulties for patient & family
  - beware of unnecessary prolongation of suffering
  - resources
When to Start Ventilation?

- often difficult to decide, depends on:
  - level of symptoms
  - patient preference / family preference
  - underlying disease progression
  - results of investigations
  - availability of resources
Decision Making Process Includes...

- Medical
- Society
- Allied Health
- Patient
- Family
- Home care
- Psychosocial support
- Financial
- Other support
Considerations - Patient & Family

- quality of life
- awareness of unnecessary prolongation of suffering
- care requirements – may need extra carers, respite etc
- effects on entire family
- difficult to make decisions to stop treatment in future
- treatment itself can be difficult to tolerate
Considerations - Medical / Allied Health

- individual’s experiences and views
- time
- care requirements
- emotional impact / spiritual / cultural views
- previous support, education, knowledge
Considerations - Society

- availability of care requirements
- cost of treatment & care
- family resources - who supports them
  - financial
  - psychosocial
- implications for family members in the future
The Options...

• Ventilation vs. no ventilation
  
  *Ventilation is not appropriate for everyone*

  • patient does not want it
  • “medically futile” – progressive disease, other

  • ideally treatment is with noninvasive ventilation (NIV)
    • relative ease of use: patients, carers, staff
    • less complications
    • more acceptable / less change in current state
The Options...

• But in some, nasal ventilation (NIV) is not possible
  • failure
    • not controlling symptoms
    • NIV not tolerated
    • Secretions may be a significant problem
  • weak upper airway muscles
  • ventilation required > 16 hours per day
• patient already has a tracheostomy for another reason
• should patients receive long term invasive ventilation???
The Dying Process

- in some - may be a definite decision to stop ventilation – this can be a very difficult & emotional time
- may decide not to treat certain life threatening problems
- medication available to control symptoms so there is not significant distress
- patient and family require lots of support
Common Pitfalls

- failure to plan
  - need to discuss early and revisit regularly
  - try and make informed decisions prior to a crisis
- unclear patient preferences – advance care planning
- insufficient time allowed for discussion, revisiting issues
- not providing adequate information re disease, & likely progression
- issues around stopping treatment and the dying process are not addressed properly