Home Mechanical ventilation in the Netherlands

View from the caregiver

University Medical Centre Groningen
Home mechanical ventilation

- Organisation in the Netherlands
  - Type of patients selected for HMV
  - Effects of HMV
- Specific problems with ALS
Total number of patients

Netherlands October 2007
Diagnoses

Netherlands October 2007
Home Mechanical ventilation Groningen

Duiverman Respir Med. 2006;100:56-65
Type of ventilatory support

Netherlands October 2007
Where do they live?

Netherlands October 2007
Survival M. Duchenne patients after start NIPPV/TIPPV
Survival University hospital Groningen

Duiverman Respir Med. 2006;100:56-65
Home mechanical ventilation

• Organisation in the Netherlands
  – Type of patients selected for HMV
  – Effects of HMV

• Specific problems with ALS
Growth of ALS on HMV

- 2001: 5% ALS, 95% Totaal
- 2007: 10% ALS, 90% Totaal
Case

Man 67 years

History 2004 april : ALS

2004 juni

tired / shortness of breath / works 4 for hours on 3 days / sleeps well / no problems to lie flat / no headache / lost 8 kg in 4 months
Case

- Physical examination:
  
  Speaks loudly / abdominal breathing / no orthopneu

- Bloodgas: pH 7.39–$pCO_2$ 6.7–$pO_2$ 10–$Bic$ 30–sat 95
Case

What to do?
Case

July 2004: starts NIPPV
    placement of PEG

Oct 2005: very happy about NIPPV
    uses NIPPV during daytime
    very happy with PEG

April 2006: uses NIPPV for 24 hrs a day

Sept 2006: died
Chronic NIV in ALS

• Why?

• When to start?

• How?

• What to do if NIV doesn’t help anymore?

• End of life problems?
ALS and survival

Bourke. Lancet Neurology 2006;5:140-7
ALS and quality of life

Bourke  Lancet Neurology 2006;5:140-7

![Graphs showing the proportion of QoL maintained over time in different groups.](Image)

**A**

Proportion QoL maintained

- NIV: 1.00, Controls: 0.75
- Days: 0, 300, 600, 900, 1200
- Numbers at risk:
  - NIV: 22
  - Standard care: 19
- p=0.0013

**B**

Proportion QoL maintained

- Days: 0, 200, 400, 600, 800
- Numbers at risk:
  - NIV: 11
  - Standard care: 9
- p=0.0004
- p=0.26
Nutritional state

Lo Coco. Neurology 2006;67:761
Chronic NIV in ALS

• Why?

• *When to start?*

• How, where?

• What to do if NIV doesn’t help anymore?

• End of life problems
When to inform patients about HMV?

• If the diagnosis is clear?

• If longfunction gets worse (VC ↓)?

• In case of signs of hypoventilation?
When to start?

- Longfunction deteriorates (VC ↓) ?
- Signs of hypoventilation ?
- Symptoms ?
- Sufficient social support ?
Chronic NIV in ALS

- Why?
- When to start?
- How, where?
- What to do if NIV doesn’t help anymore?
- End of life problems?
Non-invasive options
Admission to initiate NIV

• Problems

  – Patients are used to an individually tailored health support which is not available in hospital

  – Patients who are quadriplegic have to go to the ICU

  – In case of bulbair ALS NIV is more difficult, do we switch to TPPV?

  – Less contact with relatives

  – Difficult to sleep in hospital
EOLUS
Initiation of HMV outside the hospital

- RCT: two groups of 30 patients
- Intervention group starts NIV at home
- Primary parameter is PaCO₂
- Secondary parameter
  - QOL questionnaires
  - Lung function
  - Telemedicine
  - Costs
Chronic NIV in ALS

• Why?
• When to start?
• How?
• What to do if nocturnal NIV doesn’t help anymore?
• How is in control of the patient?
Dutch policy for HMV in ALS

Past : No, unless ...........

Present : Yes, if ..... 

Kampelmacher NTVG 2004;148:509
Meinesz NTVG 2006;150:449
Case

2001  respiratory failure by bulbair ALS
 starts TIPPV
 placement of PEG

2003  still able to walk

2004  24 hrs TIPPV , lies in bed / chair
 completely ADL dependent

2008  died
Social and medical support team

- Partner
- Kids and partners
- GP (case manager?)
- HMV, doctor, nurse, technician
- Special team home care
- Special team to provide help for ADL
- Physician for rehabilitation purposes
- Occupational therapy
- Domestic aid
How is his quality of life?

• Can not speak anymore: he did not like to talk…
• He accepted his disease
• His wife, 2 kids and two grandchildren participated in care.
• Lots of family and friends
• Patient likes to watch television, especially soccer
• Moves to a campsite every summer for 7 weeks
Where do ALS patients live?

Groningen oct 2006
Chronic NIV in ALS

- Why?
- When to start?
- How?
- What to do if NIV doesn’t help anymore?
- *End of life problems?*
End of life problems

- Bladder does not work anymore
- Intestinal function is gone
- Eyes can not be closed
- Auto regulation system is failing, low temperature
- Number of infections increases and are difficult to resolve
- If they further deteriorate admit them to the hospital (ICU)?
- When and who is switching of the ventilator?
Conclusions HMV in ALS

• HMV in ALS increases survival and QOL

• Inform patients early........

• Good timing when to start is essential

• HMV is possible at home with a multidisciplinary team

• Even at the end of the line patients might judge their QOL as “good”
Chronic TIPPV in ALS

- Tracheotomy
- TIPPV with cuff
- Problems with swallowing; PEG
- Atelectases due to no cough function
Symptoms

- Morning headache
- Shortness of breath
- Bad sleep
- Orthopnea
- Weight loss
Studie populatie

N=41

Orthopneu
N=38

Symptomatische Hypercapnie
N=3

PaCO₂ < 6.0
N=20

PaCO₂ > 6.0
N=18

Bourke  Lancet Neurology 2006;5:140-7
# Thoracic-restriction

<table>
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<th></th>
<th>n</th>
<th>acute</th>
<th>Age</th>
<th>VC</th>
<th>PaCO₂</th>
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<td>Kypho</td>
<td>60</td>
<td>42%</td>
<td>54(13)</td>
<td>1.1(0.5)</td>
<td>7.7(1.8)</td>
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<td>Polio</td>
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<td>48%</td>
<td>49(16)</td>
<td>1.2(0.6)</td>
<td>7.4(1.4)</td>
</tr>
<tr>
<td>Misc.</td>
<td>19</td>
<td>53%</td>
<td>63(10)</td>
<td>1.1(0.4)</td>
<td>8.0(2.2)</td>
</tr>
</tbody>
</table>

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