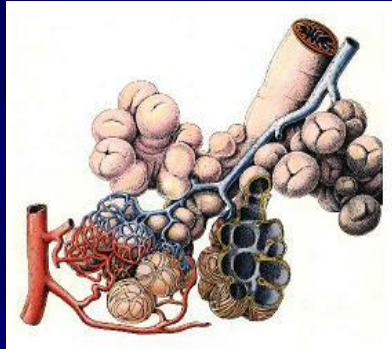


NIV in NMD- Serbian experience

**Miodrag Vukcevic
Medical School of Belgrade
CHC Bezanijska Kosa
Belgrade, Serbia**

The respiratory system

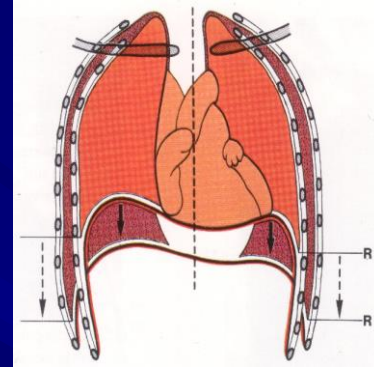
Lungs



Pulmonary failure

$\text{PaO}_2 \downarrow$ $\text{PaCO}_2 \downarrow$

Oxygen



Resp. Pump

Ventilatory failure

$\text{PaO}_2 \downarrow$ $\text{PaCO}_2 \uparrow$

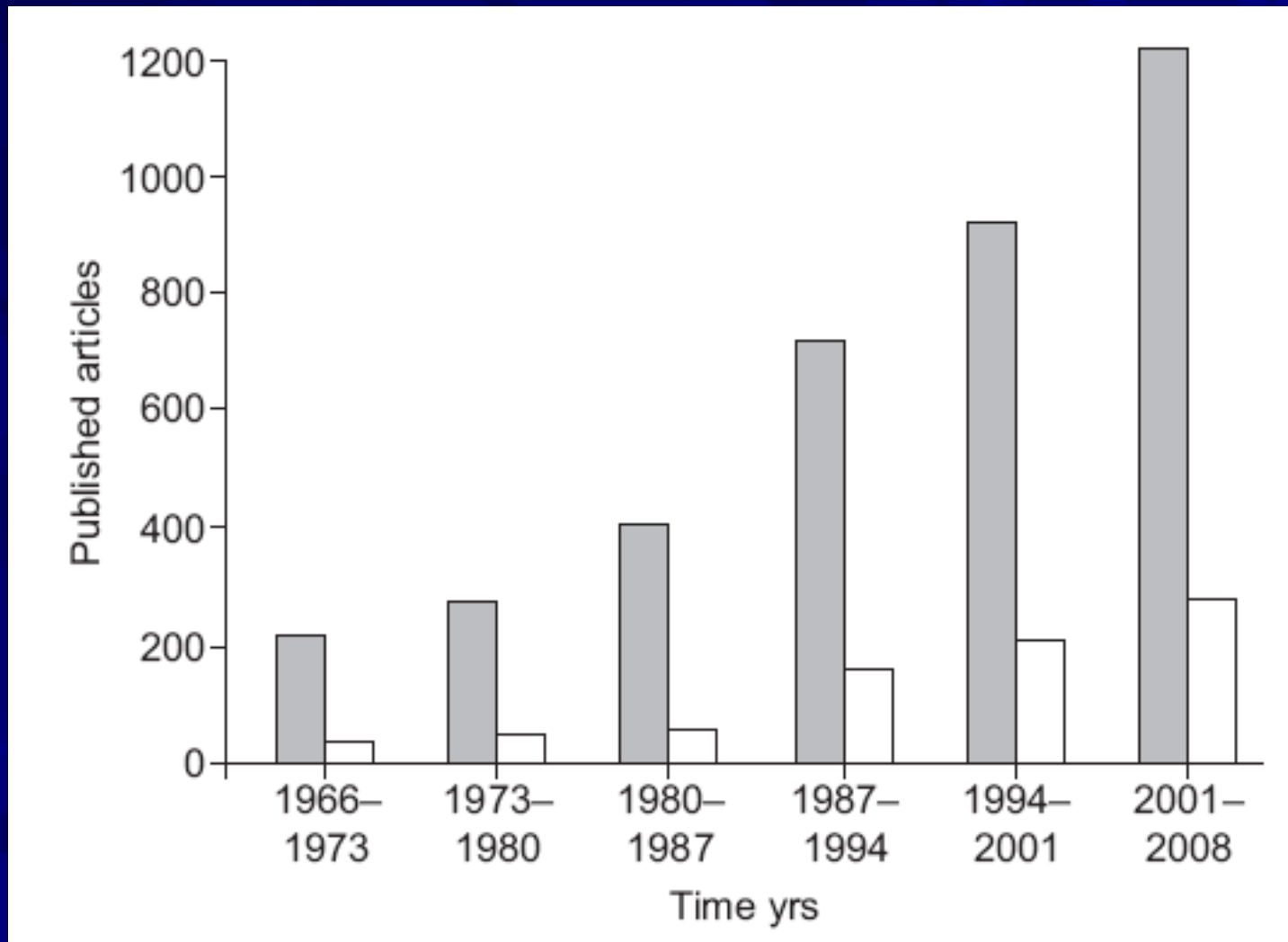
Artificial ventilation

Non-Invasive Ventilation: Definition

- Non-invasive positive pressure ventilation is any form of ventilatory support applied without the use of an endotracheal tube
- Includes:
 - CPAP with or without pressure support
 - Volume-and pressure-cycled systems
 - Proportional assist ventilation
 - Bi-level pressure ventilation

AJRCCM 2001; 163: 283 - 291

Time course of published original articles and reviews neuromuscular diseases and respiratory failure listed on PubMed from 1996–2008.



Neuromuscular diseases affecting respiratory function

■ Neuropathic disease

- Motor neuron disease
 - Amyotrophic lateral sclerosis
 - Poliomyelitis, post-polio syndrome
 - Spinal muscular atrophy
 - Paralytic rabies
- Peripheral neuropathies
 - Guillain–Barre´ syndrome, Chronic inflammatory demyelinating polyneuropathy
 - Critical illness polyneuropathy
 - Unilateral or bilateral diaphragm paralysis
 - Charcot–Marie–Tooth disease

■ Disorders of the neuromuscular junction

- Myasthenia gravis, congenital myasthenic syndrome, Lambert–Eaton myasthenic syndrome
- Botulism, poisoning with curare and organophosphate

■ Myopathies

- Acquired myopathies
 - Polymyositis, dermatomyositis
 - Critical illness myopathy
- Inherited myopathies
- Progressive muscular dystrophy
 - Duchenne muscular dystrophy
 - Becker muscular dystrophy
 - Facioscapulohumeral muscular dystrophy
 - Limb-girdle muscular dystrophy
 - Myotonic dystrophy
- Congenital myopathies
 - Nemaline myopathy, core diseases, myotubular myopathy
- Congenital muscular dystrophy
 - Ullrich congenital muscular dystrophy, Emery–Dreifuss muscular dystrophy, merosin-deficient congenital muscular dystrophy, merosin-positive congenital muscular dystrophy, rigid spine muscular dystrophy
- Metabolic myopathies
 - Mitochondrial myopathy, glycogen storage disease type 2

Background

Cohort series over the last few decades have clearly shown that **NIV increases survival, physiological outcomes and quality of life** in patients with chronic respiratory failure secondary to chest wall disease and **stable neuromuscular disorders**. Many of these individuals eg. with old poliomyelitis or idiopathic scoliosis may now live to their normal life expectancy.

Leger P, et al. *Chest* 1994;105:100-5.

Simonds AK, Elliott MW. *Thorax* 1995;50:604-9.

“No Duchenne muscular dystrophy person should ever require a trach tube or develop respiratory complications. If people follow closely what we describe, respiratory difficulties can be eliminated”

J Bach 2004!!!

➤ In inherited neuromuscular disease NIV can also extend survival eg. in Duchenne muscular dystrophy and other congenital myopathies and muscular dystrophies of childhood.

Eagle M, *Neuromusc Disord* 2002;12:926-9.

Bach JR, Ishikawa Y, Kim H. *Chest* 1998;112:1024-8.

➤ In amyotrophic lateral sclerosis/motor neurone disease (ALS/MND) NIV has been shown to reduce mortality, and improve sleep quality and energy/vitality in the face of continued motor decline.

Kleopa KA, et al. *J Neurol Sci* 1999;164:82-8.

Progression in ventilatory need

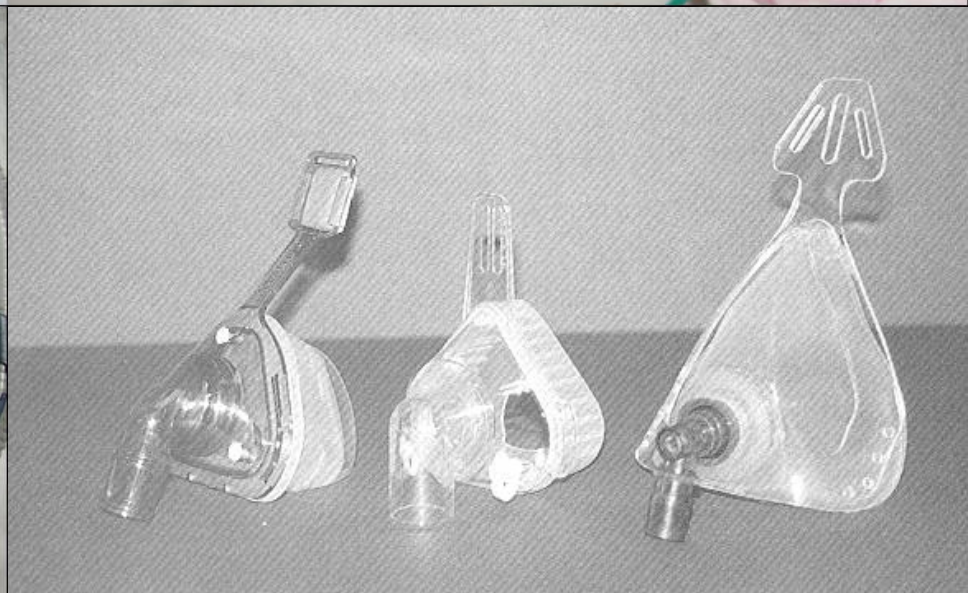
Ventilatory dependency is likely to increase over time with progressive neuromuscular disease and ventilator settings may also need to be changed.

Ventilatory adjuncts, such as mouth ventilation with a **mouthpiece** attached to a support on a wheelchair, can be helpful [Bach JR Chest 1993].

Some patients with progressive bulbar problems and/or increasing ventilator dependency **may wish to progress to invasive ventilation**. Options should be carefully discussed with the patient and family, and advanced directives formulated where possible

Interface

The crucial link between the artificial airways of the ventilatory system and the biological airways of the patient



Signs and symptoms of respiratory failure

■ Symptoms

- Increasing generalised weakness
- Dysphagia
- Dysphonia
- Dyspnoea on exertion and at rest
- Fatigue
- Sleepiness

■ Clinical signs

- Rapid shallow breathing
- Tachycardia
- Weak cough
- Staccato speech
- Accessory muscle use
- Abdominal paradox
- Orthopnoea
- Weakness of trapezius and neck muscles
- Single-breath count
- Cough after swallowing

Methods for Assessing Respiratory Muscle Weakness/Hypoventilation

- Lung volumes (VC, TLC)
- Pimax/Pemax/SNIP
- ABG: PaCO₂/ PaO₂/pH/BE
- Cough strength
- MIC
- Noninvasive blood gases (PETCO₂/PTCCO₂/Oximetry)
- Sleep studies
(Oximetry/PETCO₂/PTCCO₂/Polysomnography)
- Non-volitional tests of muscle strength

Laboratory data

- $VC \leq 15 \text{ mL/kg}$, $VC \leq 1 \text{ L}$ or 50% drop from value in stable state, or >20% drop from sitting to supine position
- Maximum inspiratory pressure $\leq 30 \text{ cmH}_2\text{O}$
- Maximum expiratory pressure $\leq 40 \text{ cmH}_2\text{O}$
- Nocturnal desaturation
- $\text{Pa,CO}_2 > 45 \text{ mmHg}$ (6.7 kPa)

Consensus Conference: Clinical Indications for NIV in CRF

Restrictive disorders

- Diagnostic certainty
- Symptoms eg. fatigue, dyspnoea, headaches
- 1 of the following:
 - $\text{PaCO}_2 > 6.0 \text{ kPa}$
 - Nocturnal $\text{SaO}_2 < 88\%$ for > 5 minutes
 - $\text{MIP} < 60 \text{ cmH}_2\text{O}$ or $\text{FVC} < 50\% \text{ pred}$ in progressive disorders

Chest 1999;116:521-524

Home mechanical ventilation in 16 European countries



Situation 2005

27.000 patients

COPD/Lungs

36 %

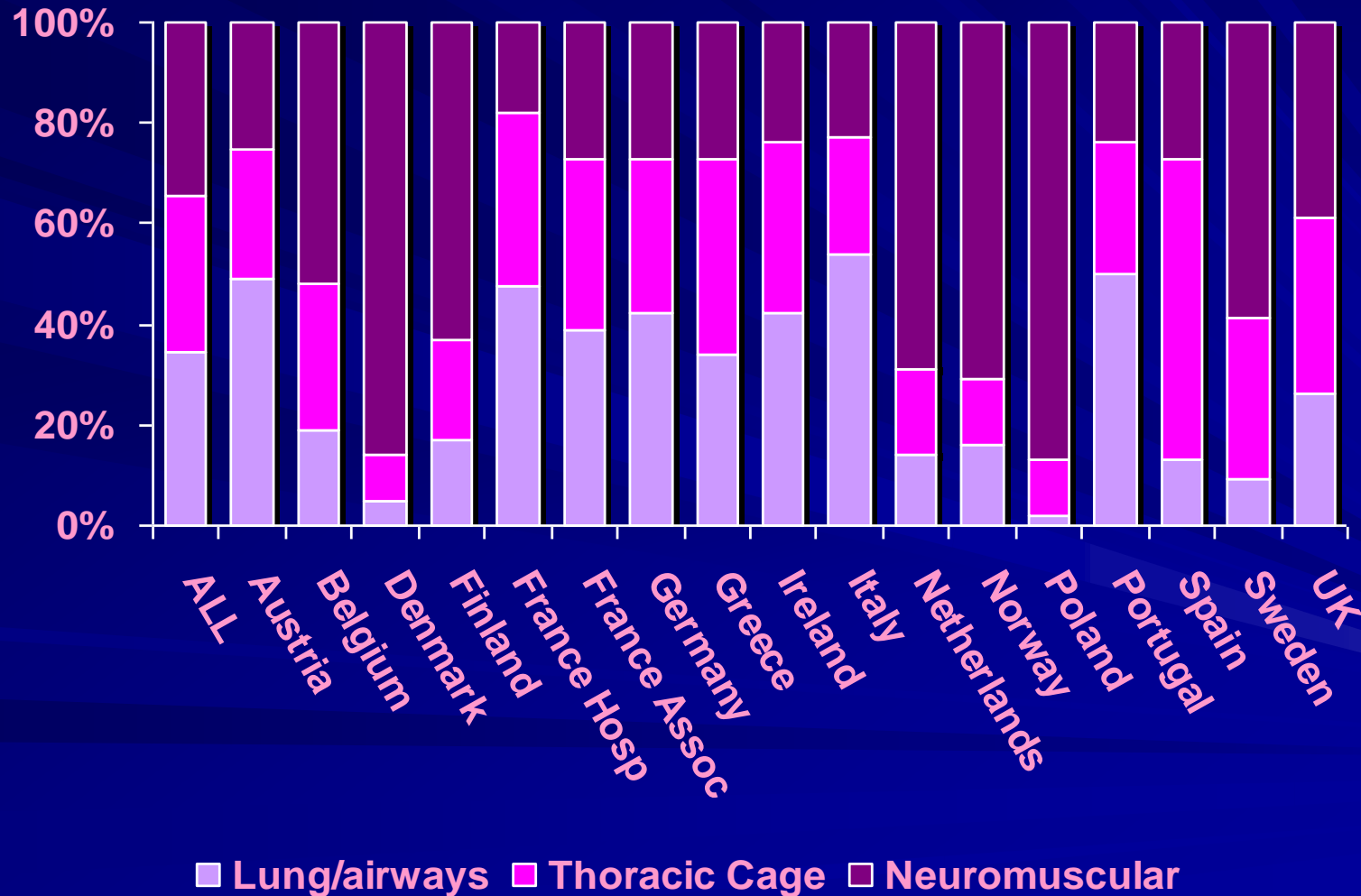
Rib cage disease

31%

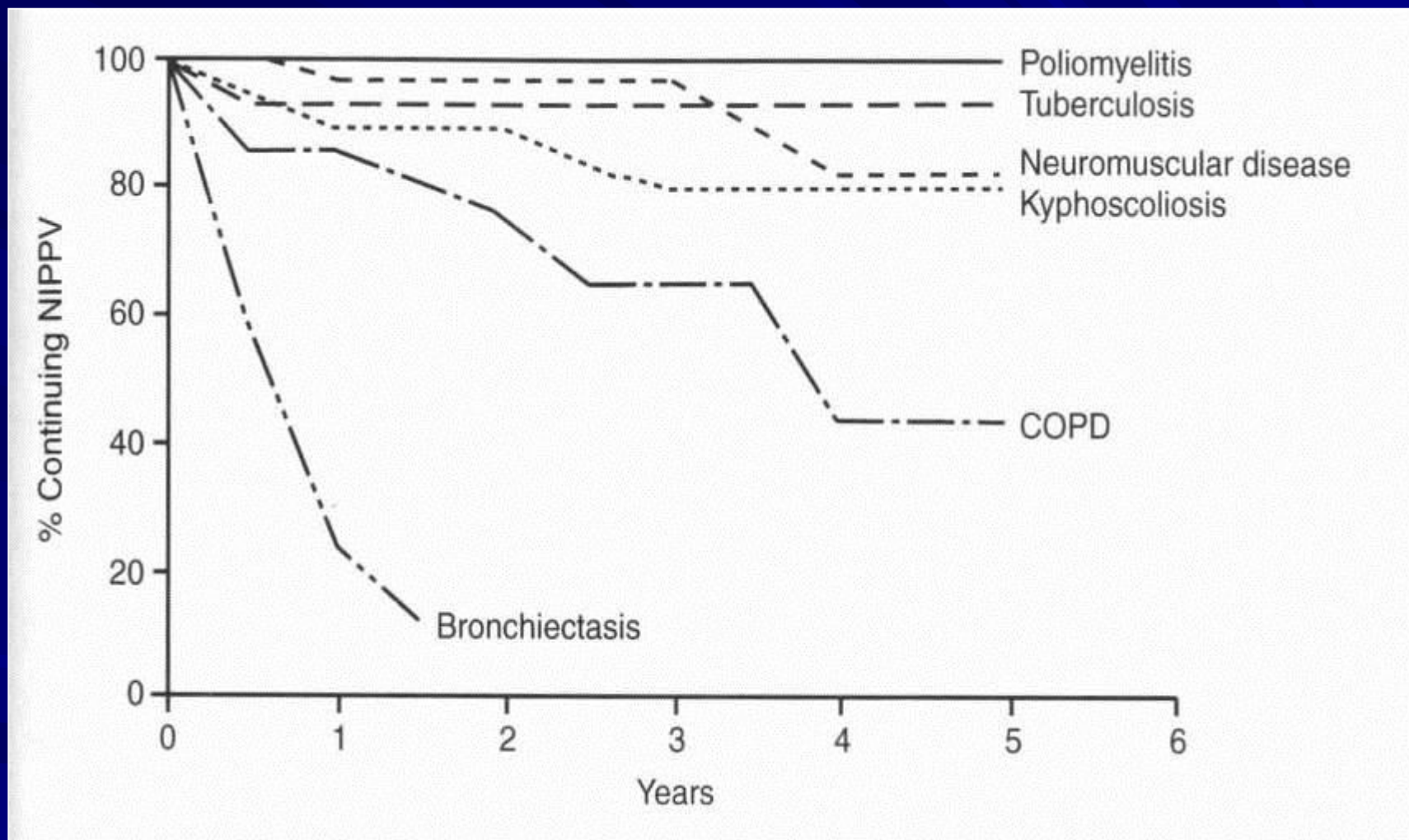
Neuromuscular

33 %

Disease Categories in Europe



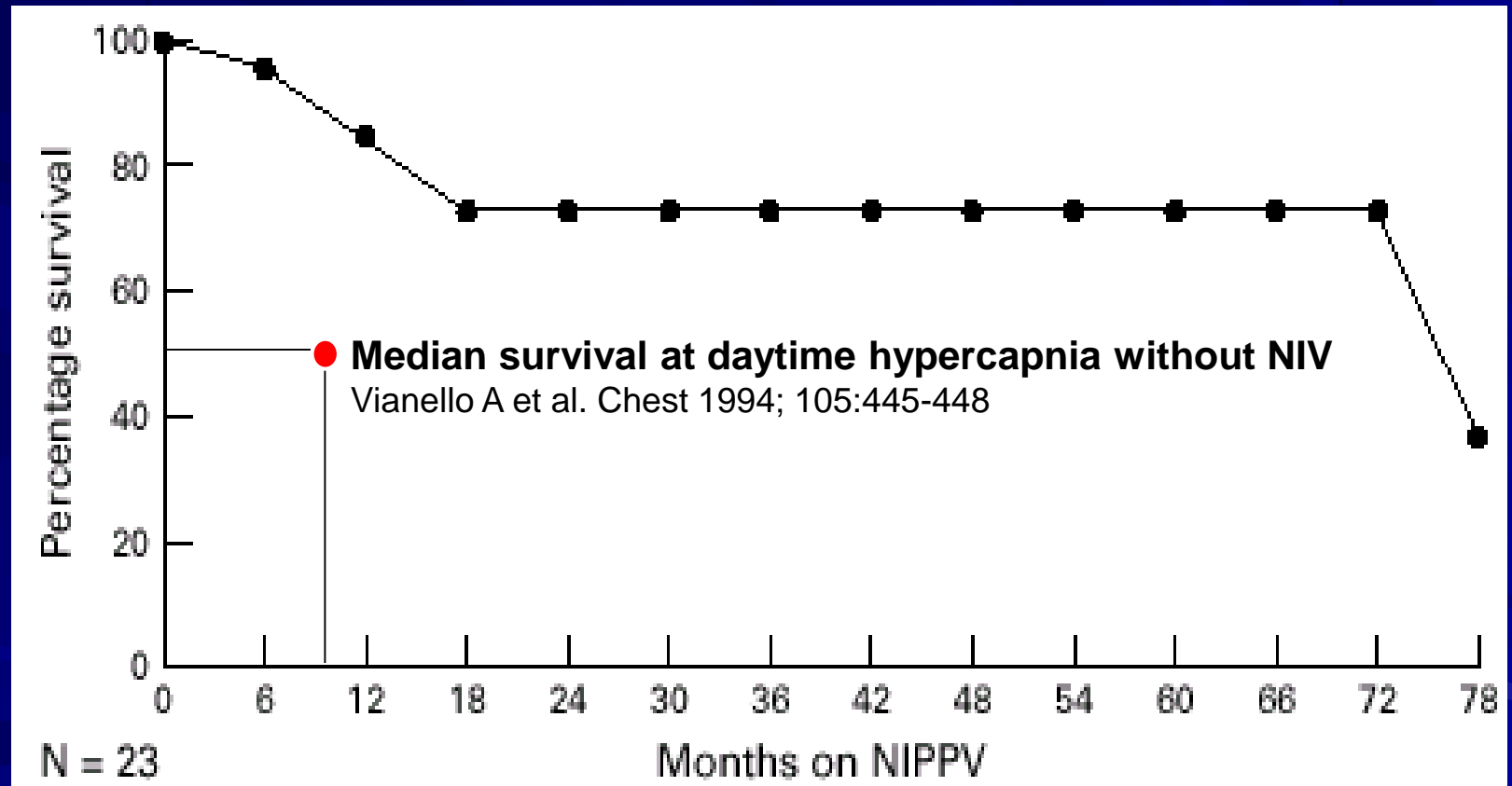
Survival: Probability of continuing domiciliary NIV



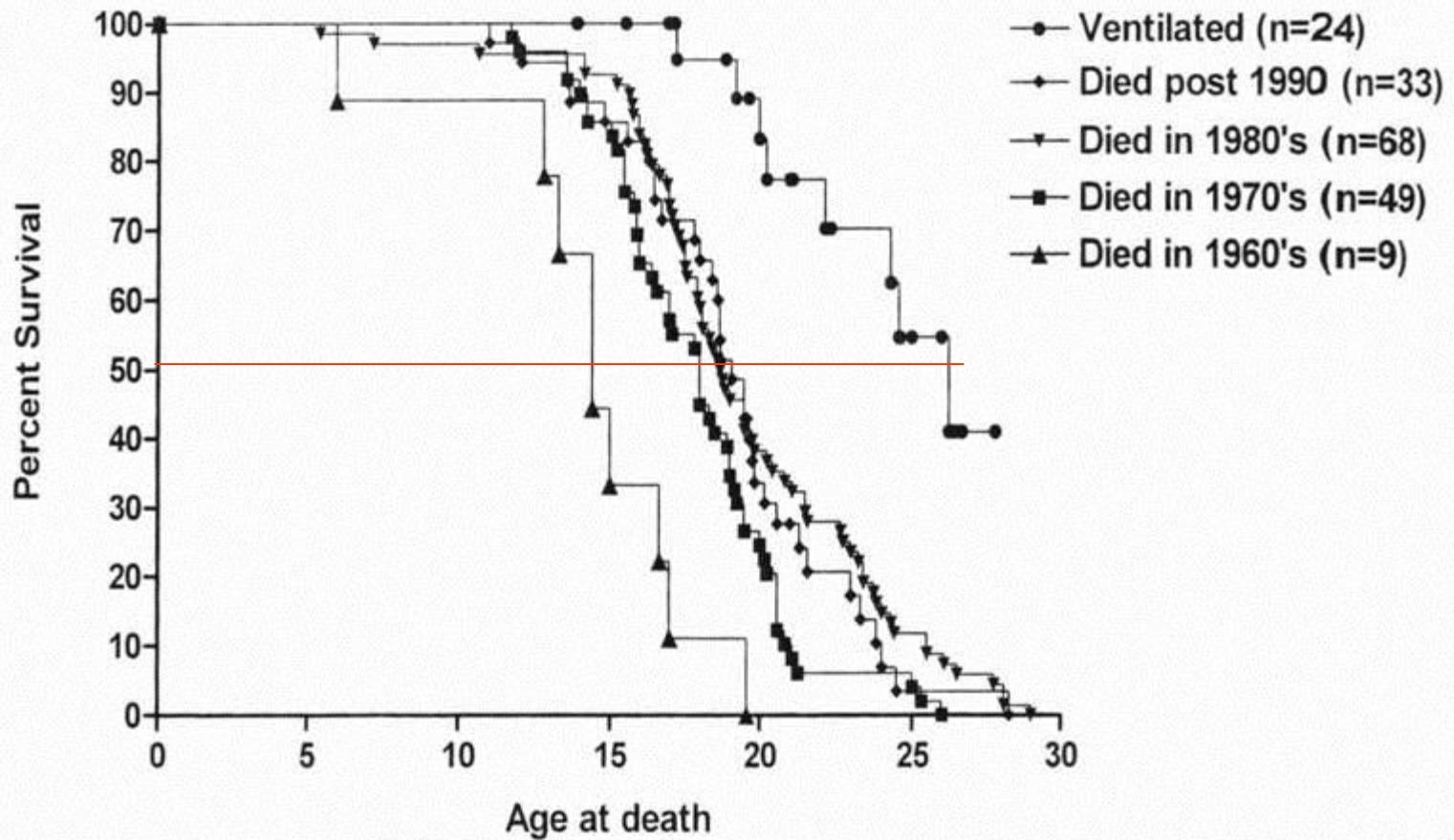
Simodns A., Thorax 1995

Impact of nasal ventilation on survival in hypercapnic Duchenne muscular dystrophy

A K Simonds, F Muntoni, S Heather, S Fielding

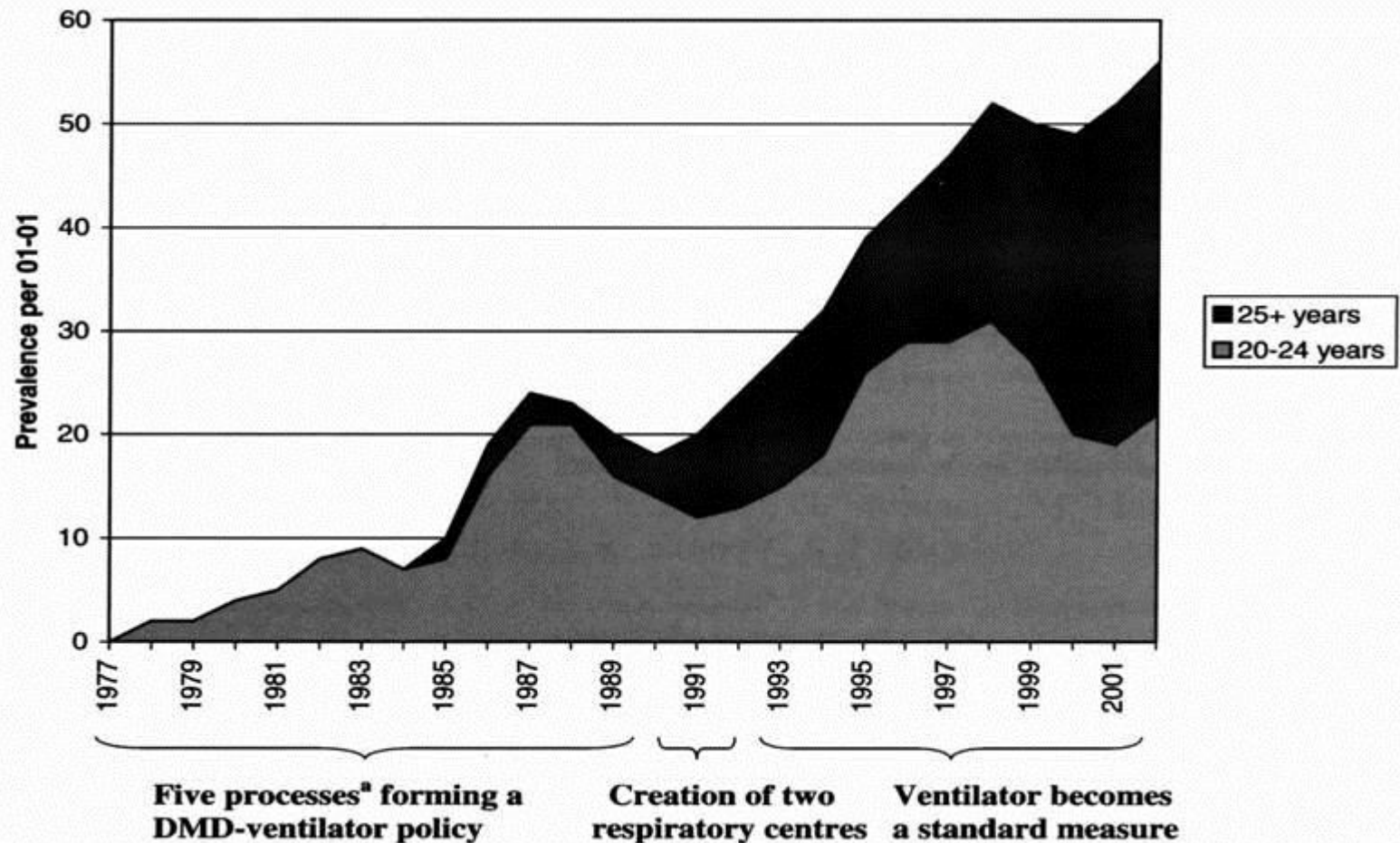


Trends in survival in Duchenne MD



Eagle et al Neuromusc Dis 2002

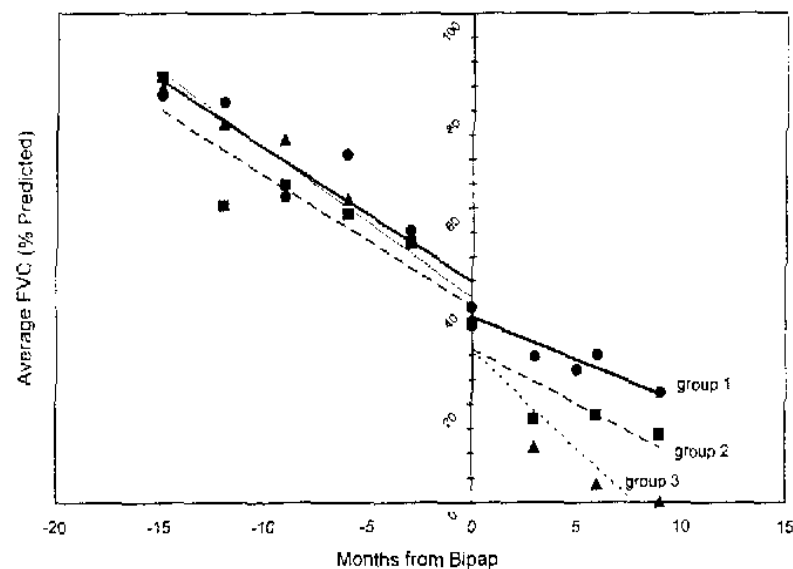
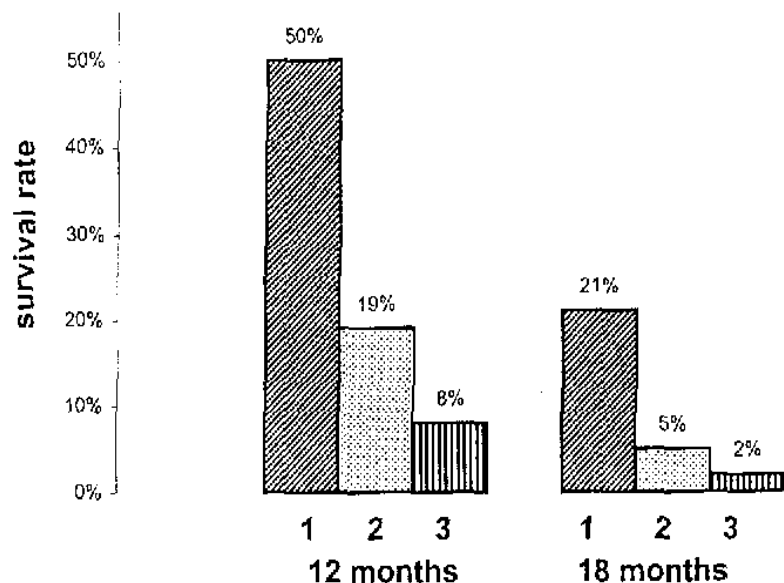
Management trends in DMD



Jeppesen J Neuromusc Dis 2003;13:804-12

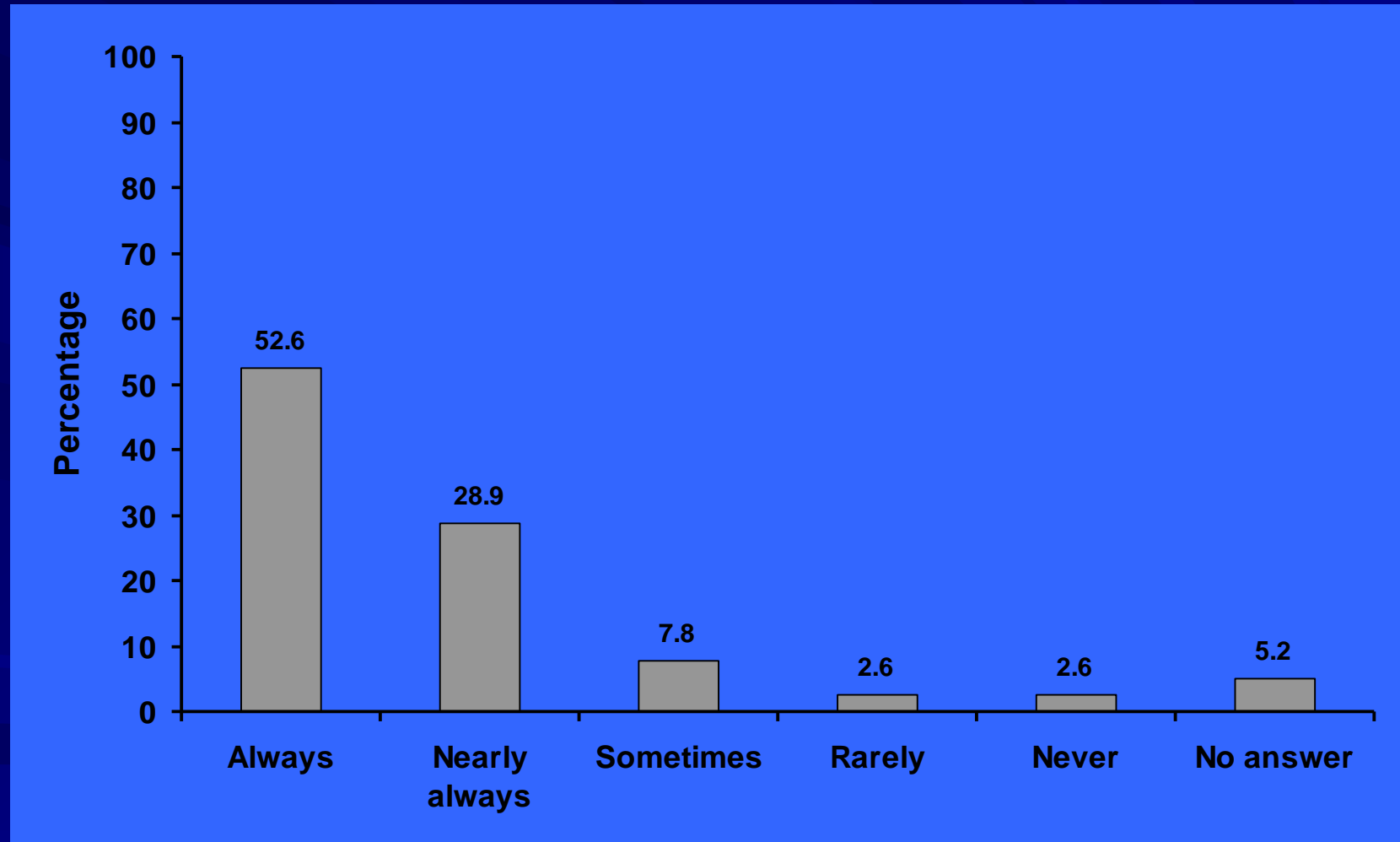
Survival and lung function in MND/ALS with NIV

Kleopa et al 1999 J Neurol Sci



	Group 1	Group 2	Group 3
<i>NIV use</i>	> 4hrs	< 4 hrs	Nil
<i>Survival</i>	Gp 1: 14.2 mths	Gp 2: 7.0 mths	Gp 3 :3.0 mths

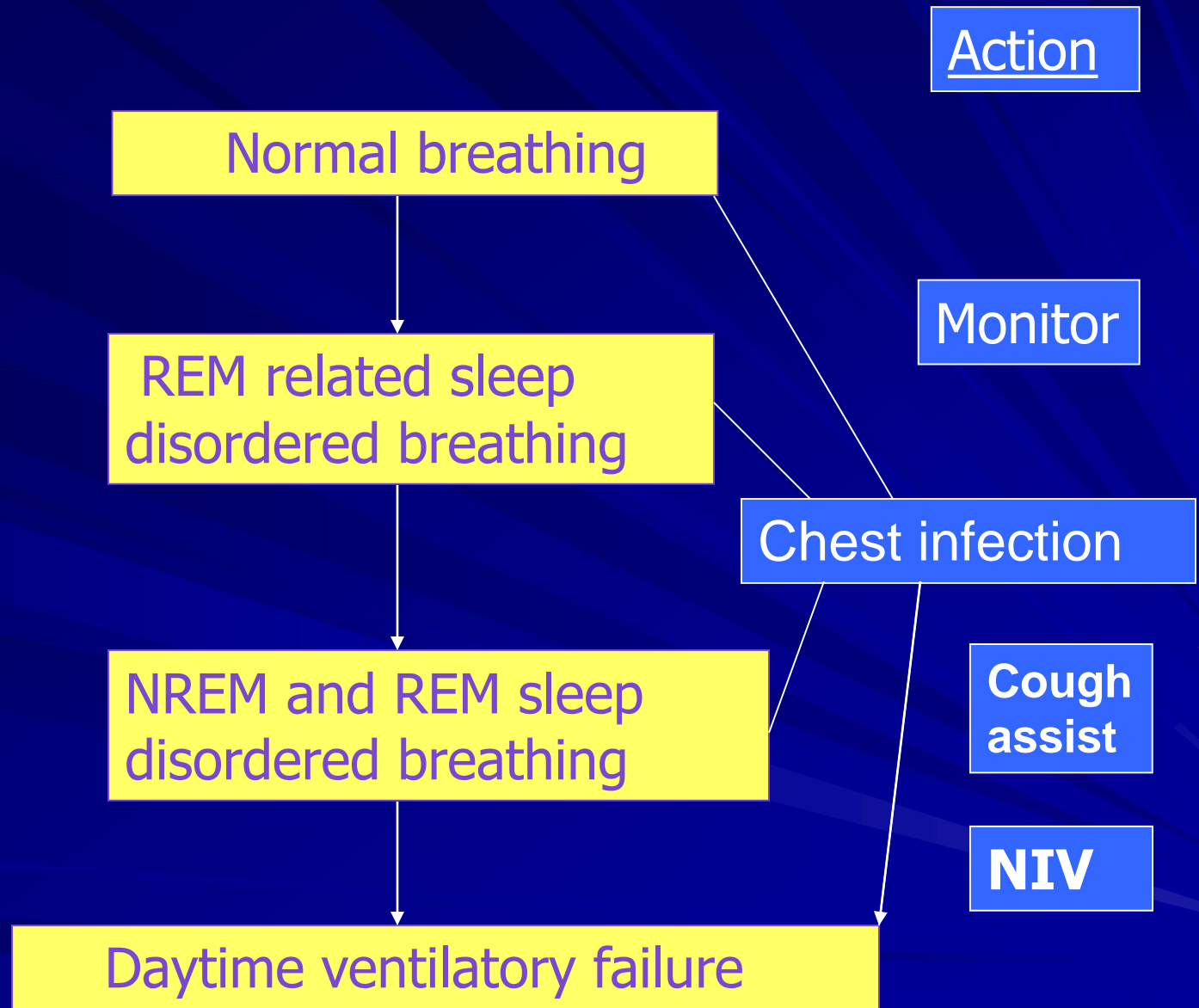
Frequency of informing families about NIV



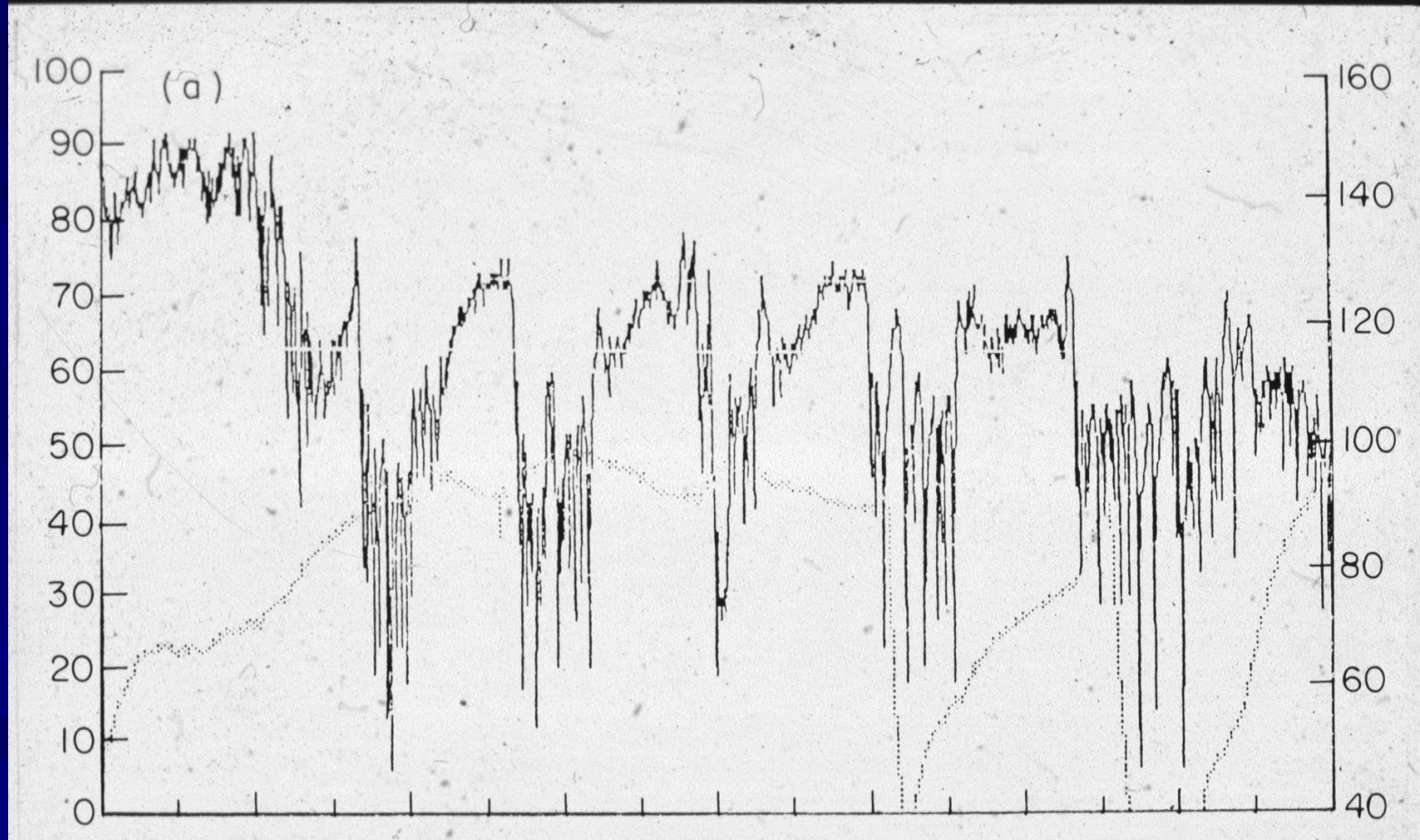
Evolution of
respiratory
failure

FVC <
40% pred

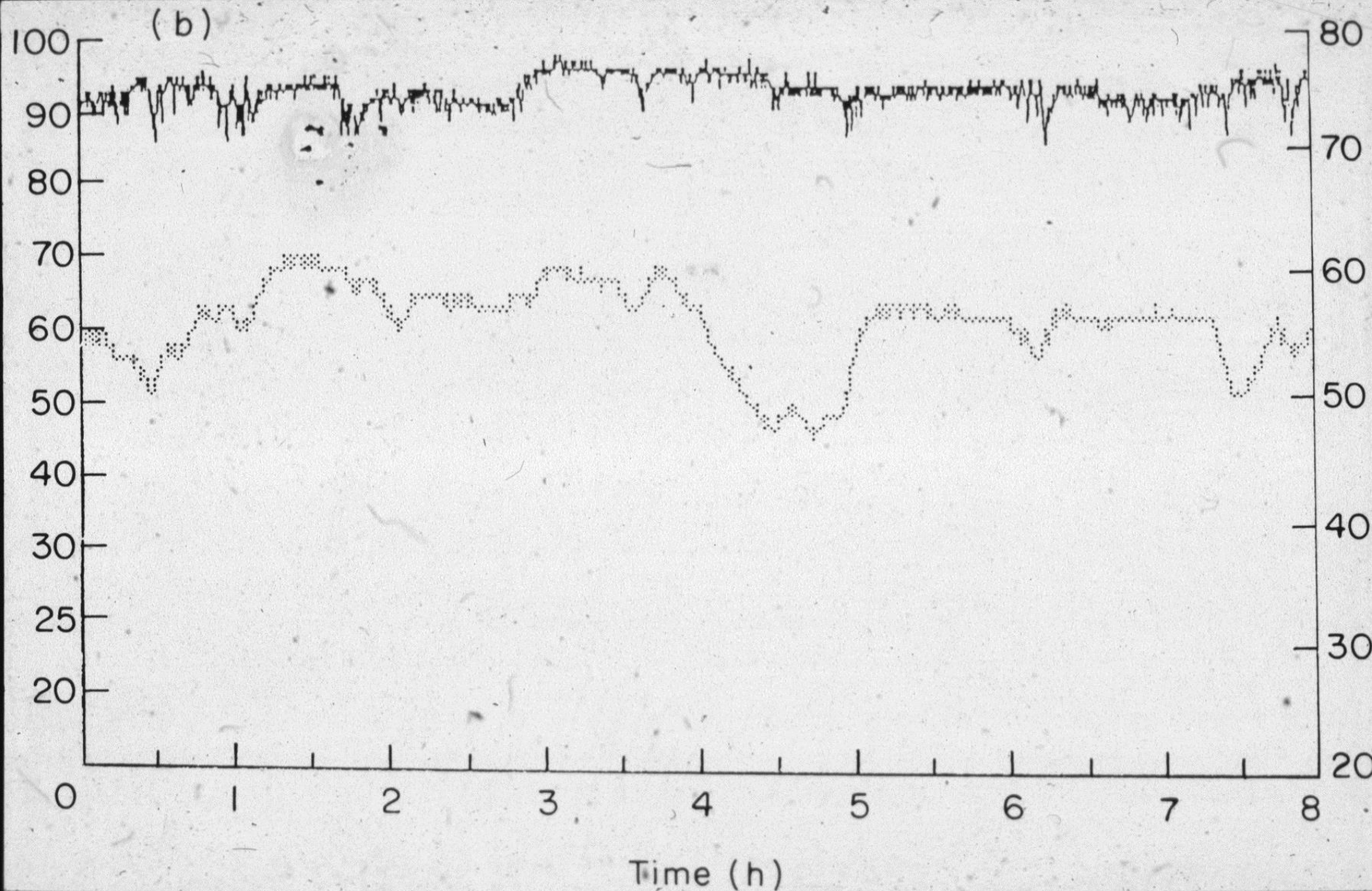
FVC <
20% pred



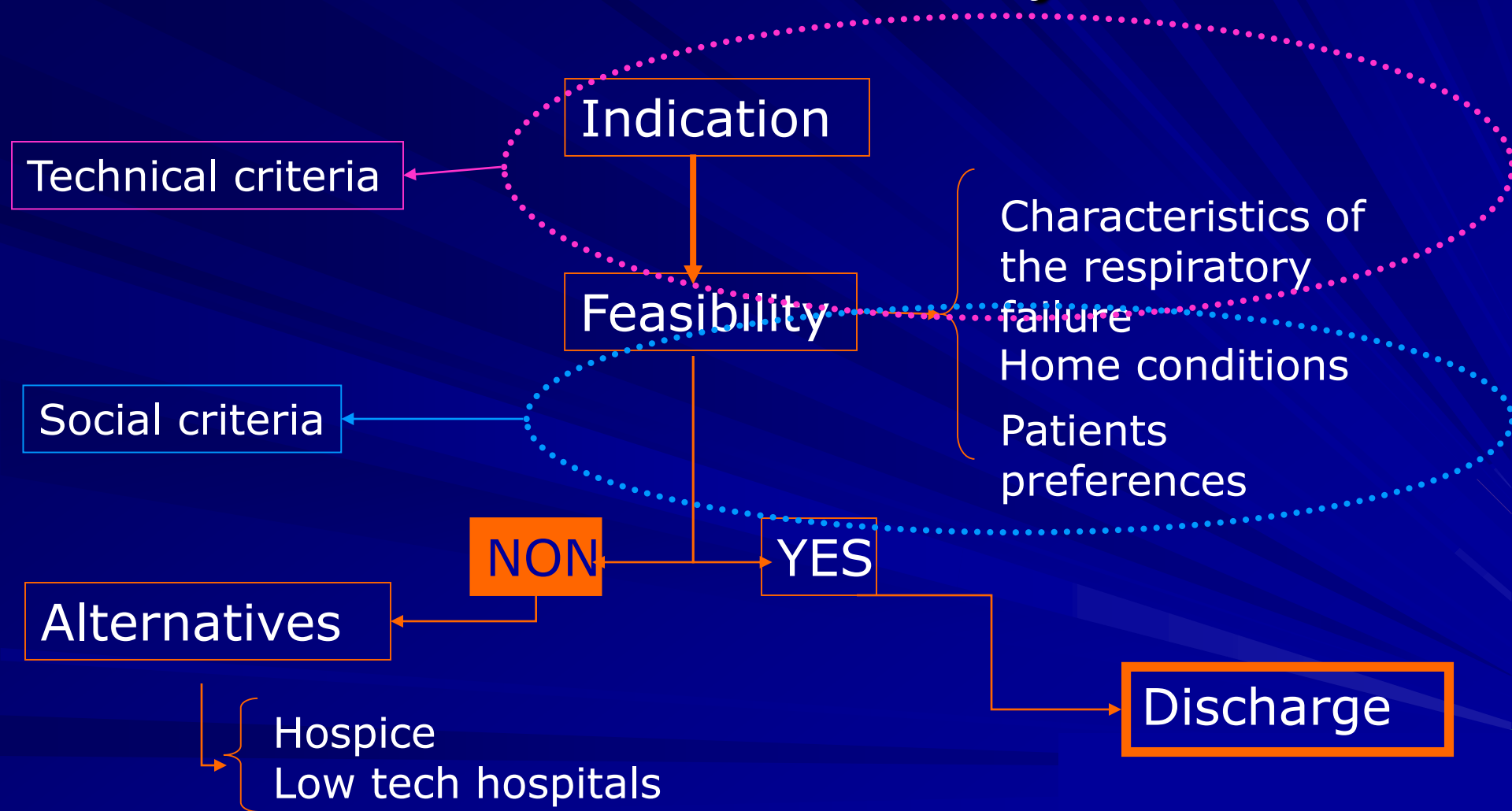
Oxymetry/Transcutaneous CO₂ before NIV



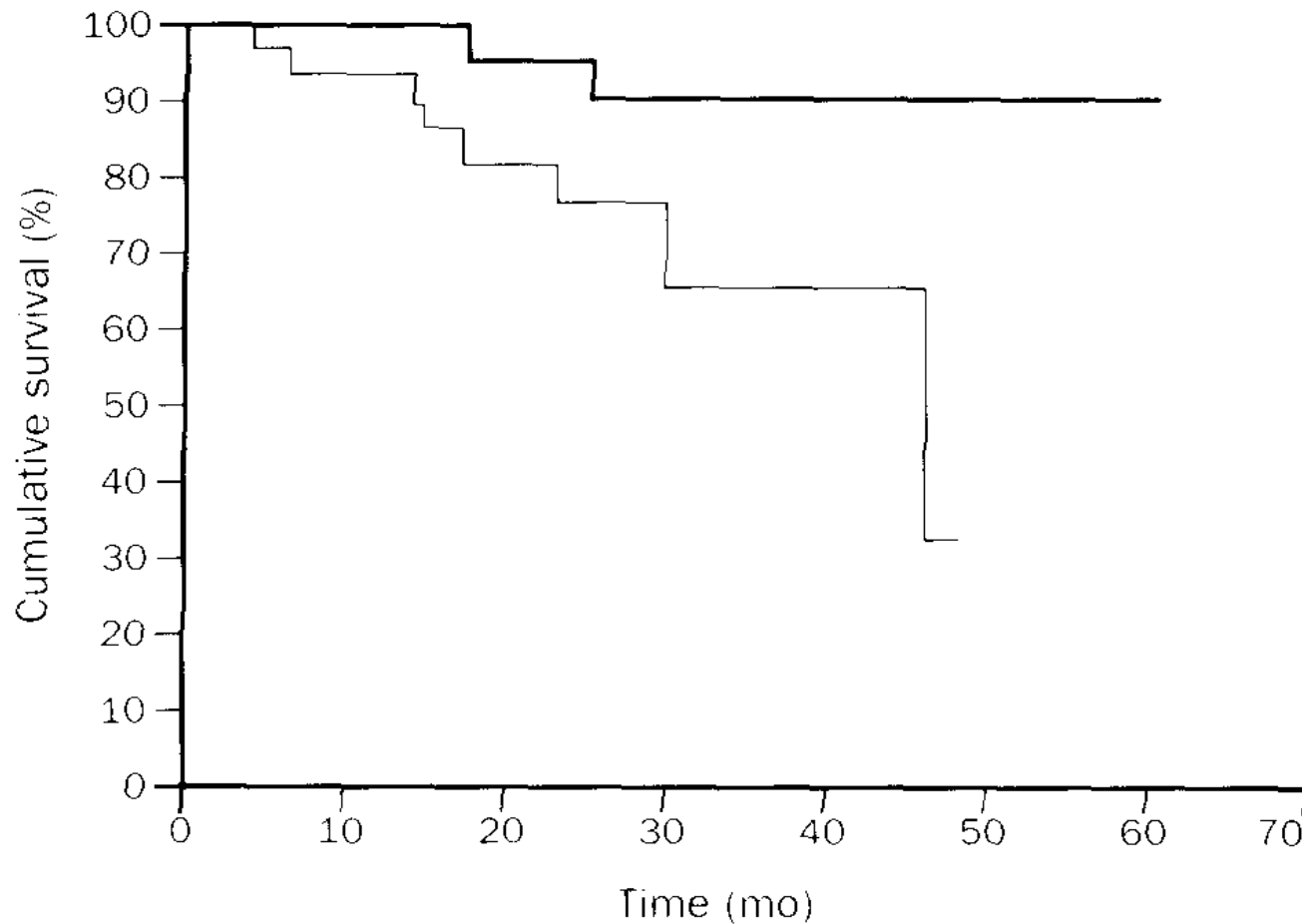
Oxymetry/Transcutaneous CO₂ during NIV



NIV: Feasibility



Prophylactic NIV in Duchenne MD



Kaplan-Meier plot of survival from randomisation in DMD patients receiving nocturnal NIPPV or conventional treatment

Raphael et al Lancet 1994

Ethical issues regarding NIV

- Patient autonomy.
- Information, education
- Continuum of care
- Advanced directives.

**Patients should identify limits of the therapy,
especially regarding tracheotomy**

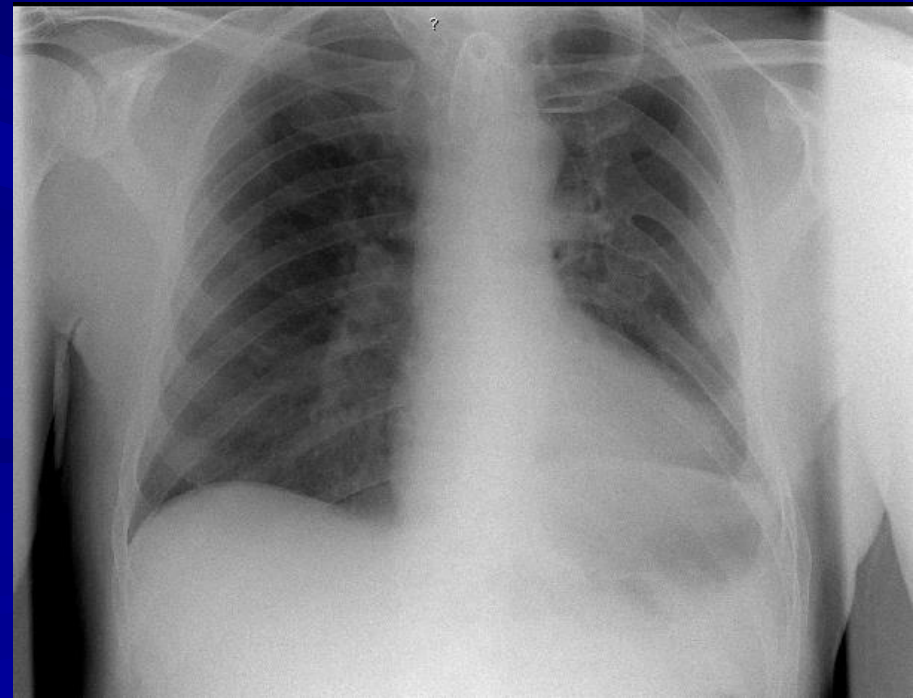
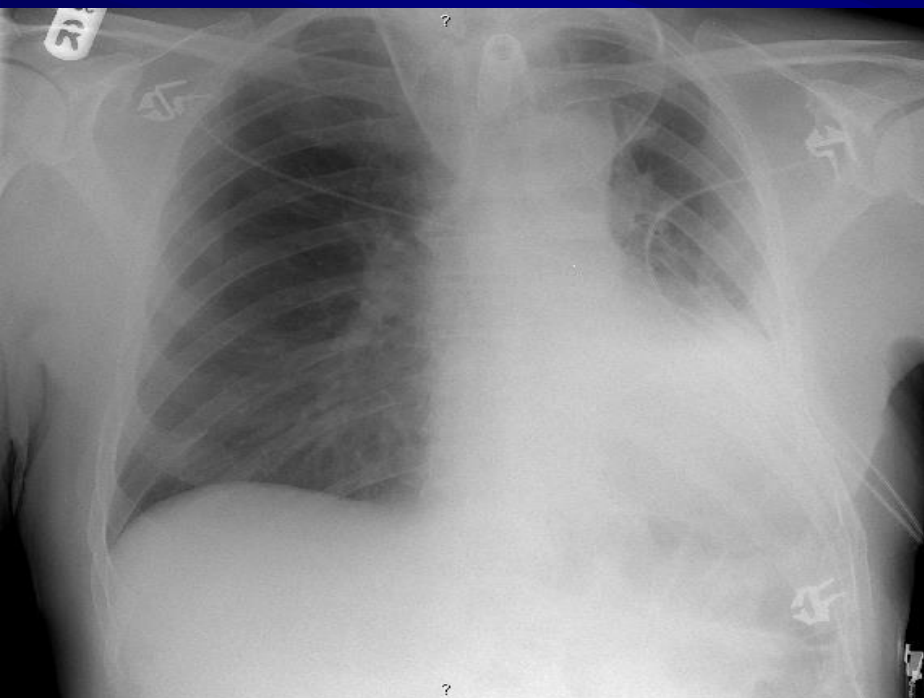
Patients can change their mind (or directives) at any time

NIV at the end of life

- It's not easy to decide whether the burdens exceed the benefits in NIV at the end of life.
- The risk of NIV is to prolong an inevitable course towards death.
- We deal with two main situations:
 - To initiate NIV late in the course of the disease
 - Withdrawal NIV in a long-term user

Equipment needs for NIV

- Respiratory accessories
 - **Humidification**
 - **Oxygen supplementation**
 - **Drugs nebulisation**
 - **Power supply: battery power source, backup ventilator**
- Monitoring
- Secretions management
- Daily living activities
- Communication



Patient
with
NMD

Respiratory
muscle
weakness

NIV

**To increase efficacy NIV may need to
be used in conjunction with other
airway clearance techniques**

Decreased
ability to
take a deep
breath in

Decreased
ability to
cough

Decreased
chest wall
compliance

Recurrent
respiratory
tract
infections

NIV

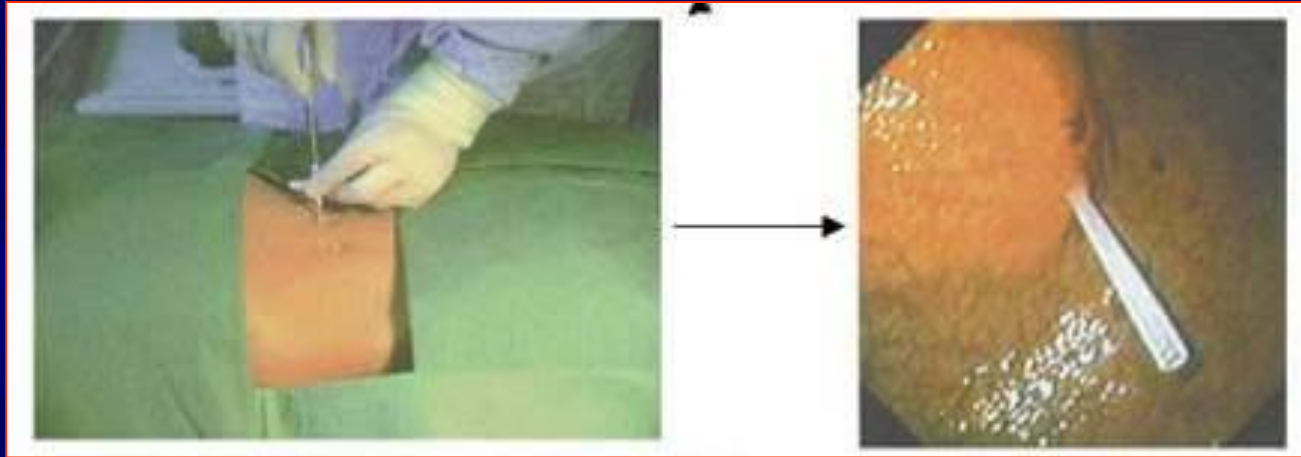
NIV

NIV

NIV

Noninvasive ventilation allows gastrostomy tube placement in patients with advanced ALS

Neurology. 2001; 56:413-4



The use of NIV for ventilatory support during percutaneous endoscopic gastrostomy (PEG) tube placement is described in five patients with advanced ALS, four having significant bulbar symptoms.

No respiratory complications occurred in any of these patients, who were considered to be at high risk for PEG placement

National referral center

- NIV started 2004 in Serbia
- Organized during 2007/2008.
- 250 PV403 units/ViVo 40

Actual situation

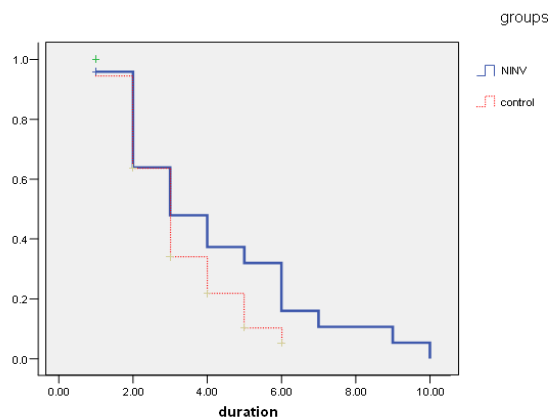
Serbia	
Population	8 million
Reimbursement for home ventilators (yes/no)	Yes
Year started	2003
Type of ventilator reimbursed (life support, pressure support, both)	both
Initiatives to start home ventilation -societies involved	Yes
Specialized center for long-term ventilation	Yes
Society formed (yes/no) -members	Yes

Actual situation

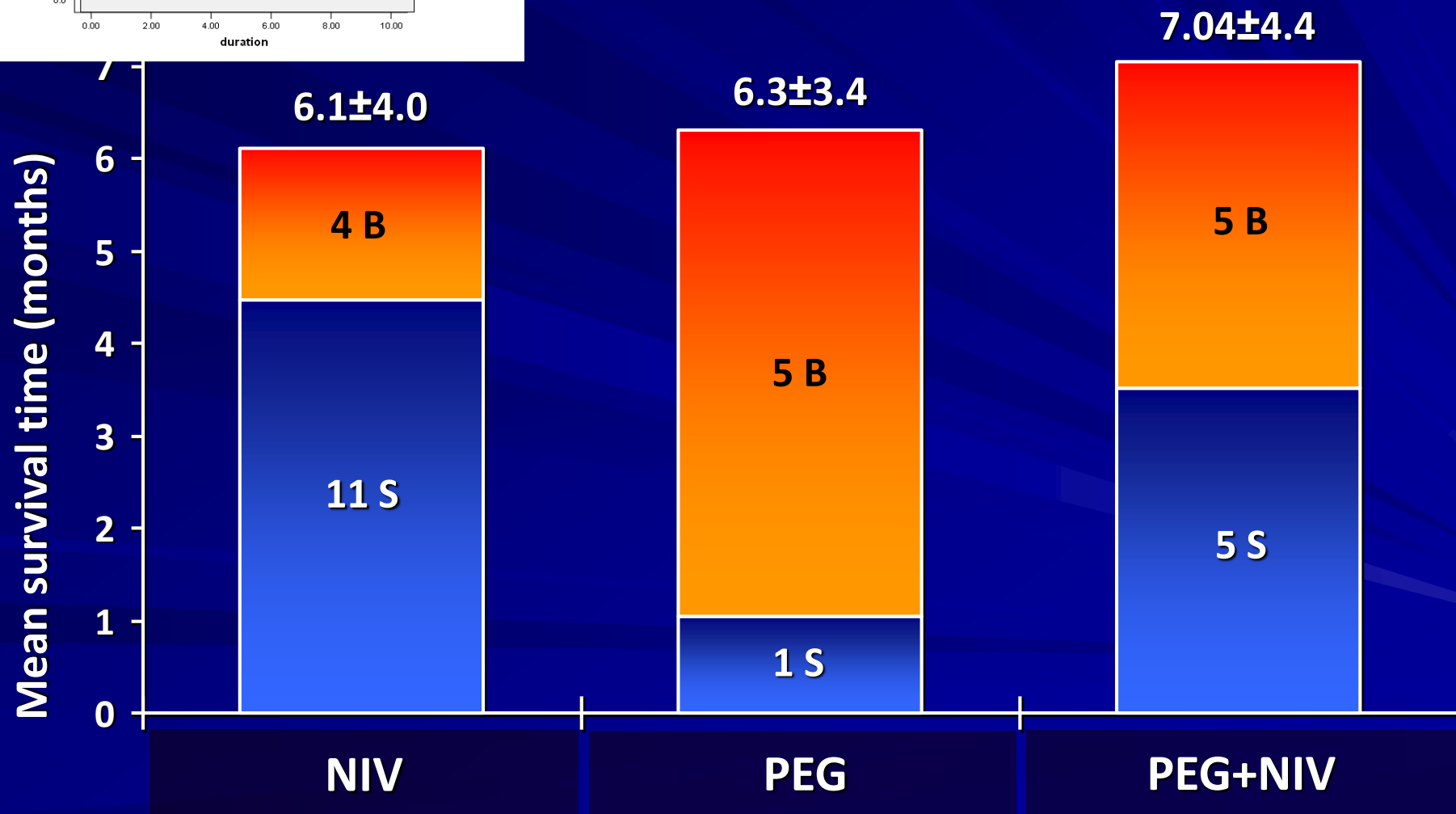
Serbia

Total number of hospitals	30
Number of hospitals involved in home-care ventilation	3
-Adult ICU	?
-Pediatric ICU	5
-Pulmonary department (incl. ICU)	24 (3)
-Neurology department (incl. ICU)	24 (?)
Number of ventilator users	250 (NMD), 60-80 OHS, 5 Kyph, 32 chi
- Invasive	10% (2007)
- Noninvasive	240 (2004)

Cumulative Survival



The mean survival time in three subgroups after NIV, PEG, NIV+PEG



ALS patients with NIV and PEG



Conclusion

- It is very important to think about how to organise the care of patients on HMOV: it must seek a balance between the role of reference centres and accessibility to local hospitals.
- Networking should be a reasonable alternative.

Conclusion

- Prolonging survival by many years and improving QoL in a previously lethal condition should be considered as a major progress in medicine.
- Even when survival is not prolonged, such as in ALS patients with severe bulbar involvement, NIV therapy may still improve QoL.
- Having said that we must remember that no care is possible without the sacrifice of the patient's family and caregivers.

Crucial final considerations

- Demonstrate the **effectiveness of NIV in your hospital** (ideally use in patients with better chance of success first) to overcome resistance from some colleagues eg. anaesthetists.
- Vigorously **lobby health authorities for purchase** of first few ventilators: highlight likely cost savings.
- **Present results of successful use** at local and national meetings, ask for NIV to be incorporated in national guidelines, publish results.
- Energise **nursing team, respiratory physiotherapists, students.**
- Show colleagues how NIV can help them: intensivists with weaning; neurologists with NIV for ALS, DMD; cardiologists with NIV for acute pulmonary oedema
- Inform the media you have a new treatment possibility that is **lifesaving**. Give newspaper interviews, write articles
- **Encourage providers to support your endeavours**: tell them investing in you will yield increased use of their product
- If you work in a system with **very low health resources**, it helps to be able to **repair equipment**. With **safe cleaning & common sense much 'disposable' equipment can be reused**



A good death

- Death is an important part of life.
- Sometimes death is seen as a failure.
 - In that case medicine does not give the attention to helping people to die a good death.
- **Learning from palliative medicine, every physician should care for their patients in the end of life period.**