MANAGING RESPIRATORY PROBLEMS IN PEOPLE WITH NMDS

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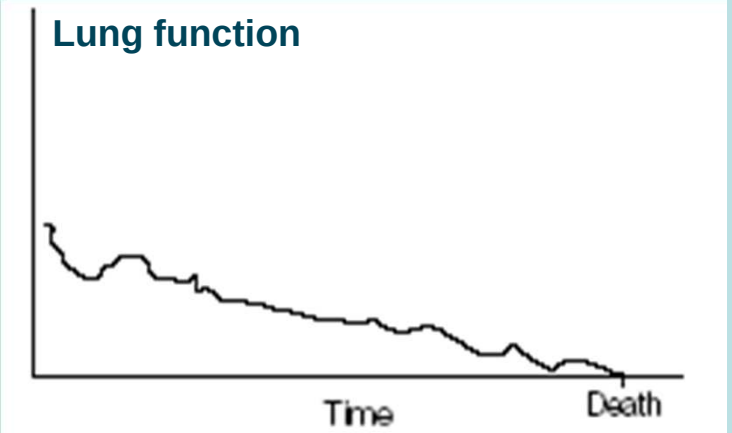
Probability of Respiratory Failure

- Inevitable: Duchenne muscular dystrophy
  Type I Spinal muscular atrophy (SMA)
  Motor Neuron Disease (MND-ALS)
- Frequent: Limb girdle MD 2C,2D,2F,2I
  Nemaline myopathy
  Int SMA
  Acid maltase deficiency
  X linked myotubular myopathy
  Ullrich congenital muscular dystrophy
  Congenital myasthenia
  Congenital myotonic dystrophy
- Occasional: Emery Dreifuss MD, Becker MD, Bethlem myopathy, Minicore, central core myopathy
- Uncommon: Facioscapulohumeral MD, Mitochondrial myopathy, Limb girdle MD 1, 2A,B,G,H, Oculopharyngeal muscular dystrophy

Course of Respiratory Failure in NMD

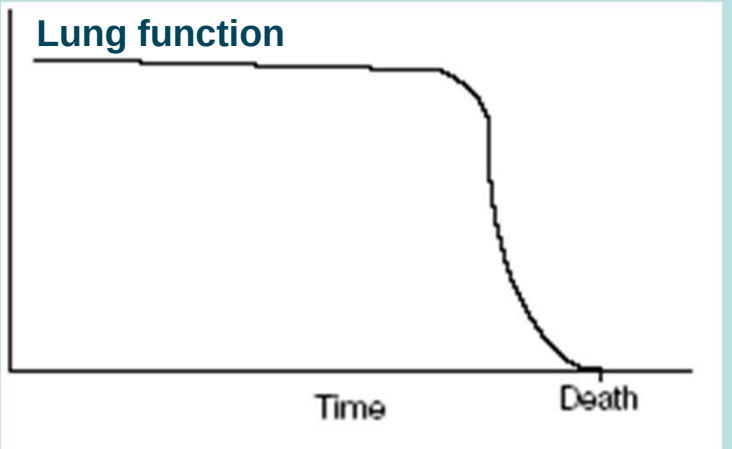
Trajectory 1: Progressive RF

A progressive, reasonably predictable RF developing over a period of months, or, in some cases, years.

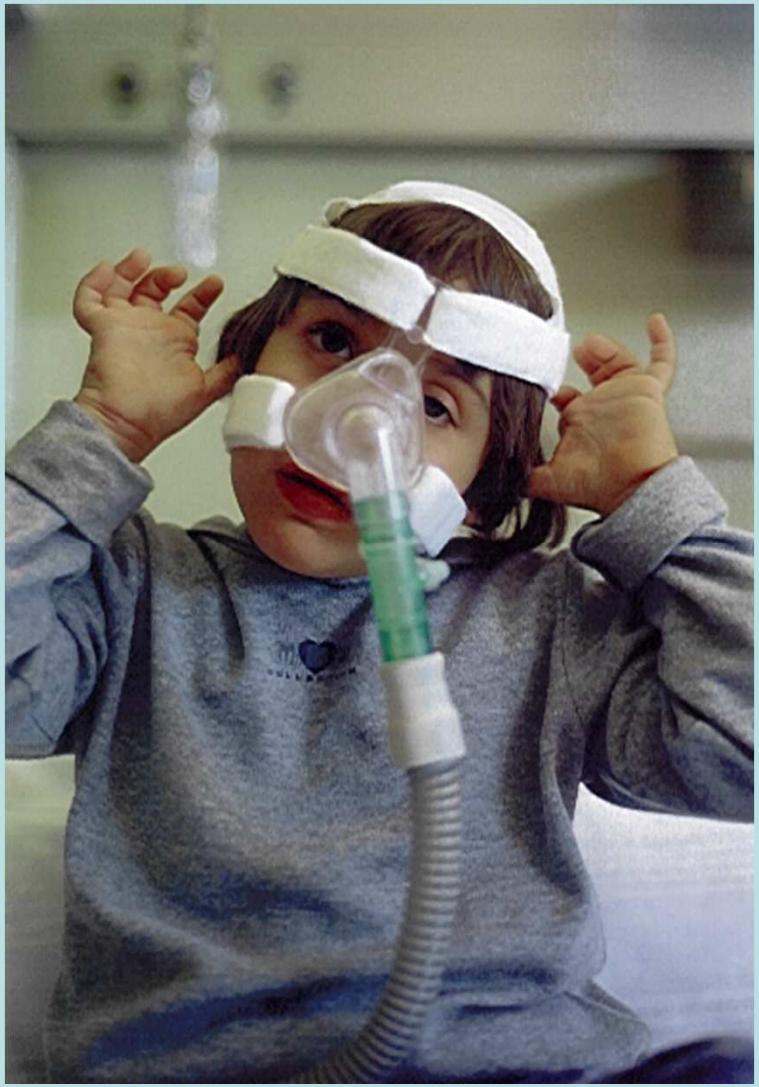


Trajectory 2: Acute RF

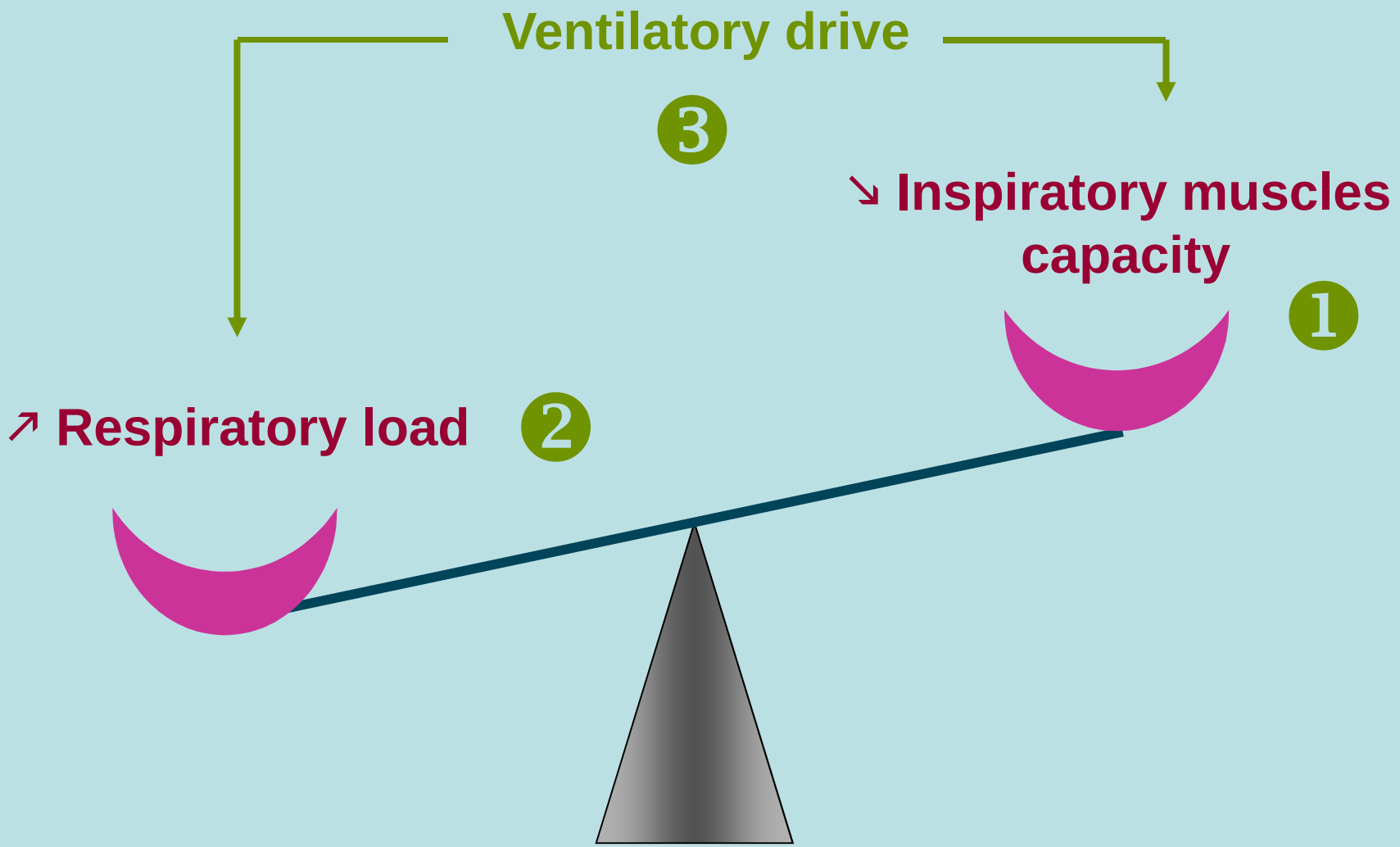
An unpredictable, acute, often severe RF requiring admission to hospital and intensive treatment.



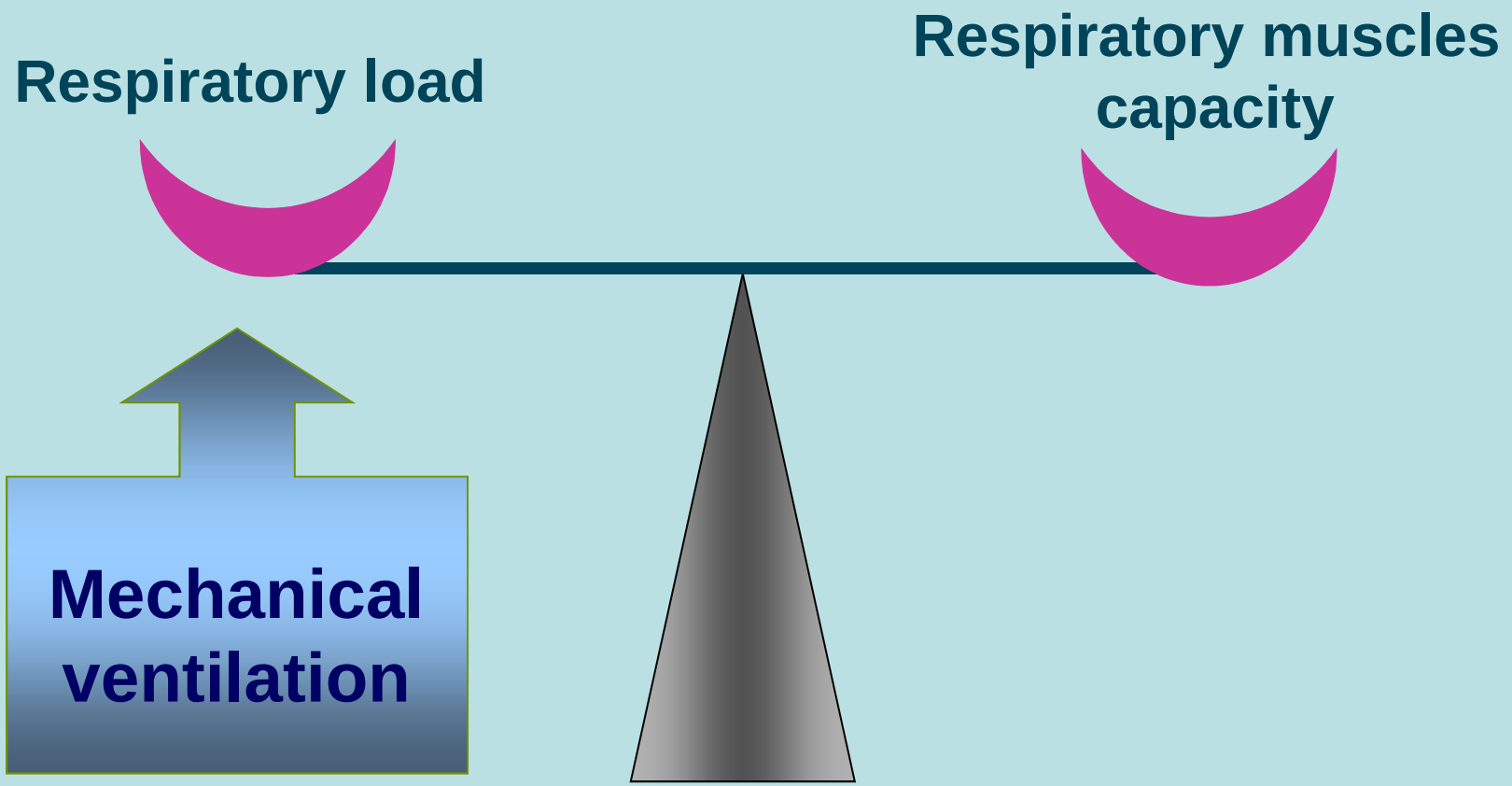
Management of Slowly Progressive RF by Long-Term NIV



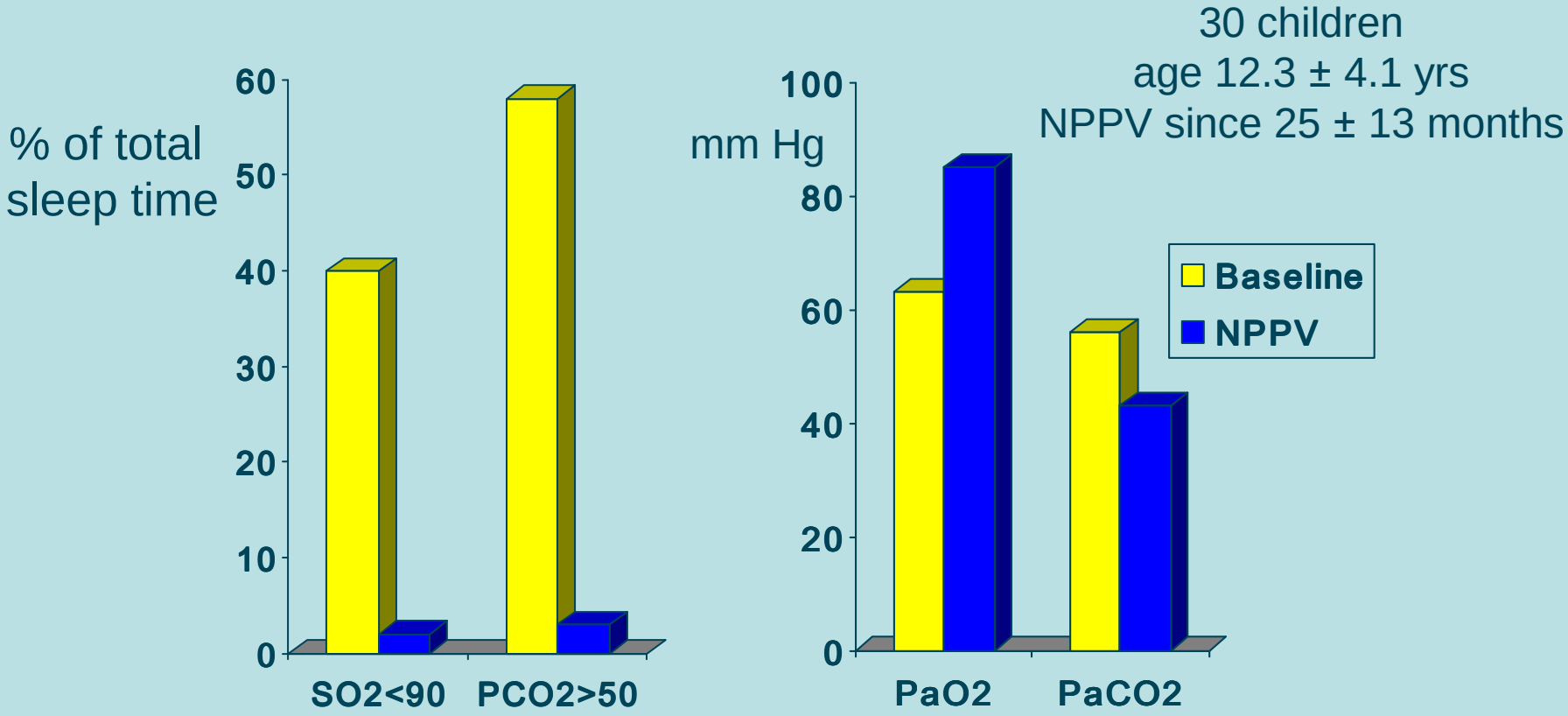
Ventilatory imbalance in patients with muscular dystrophy



Mechanical ventilation restores the ventilatory balance

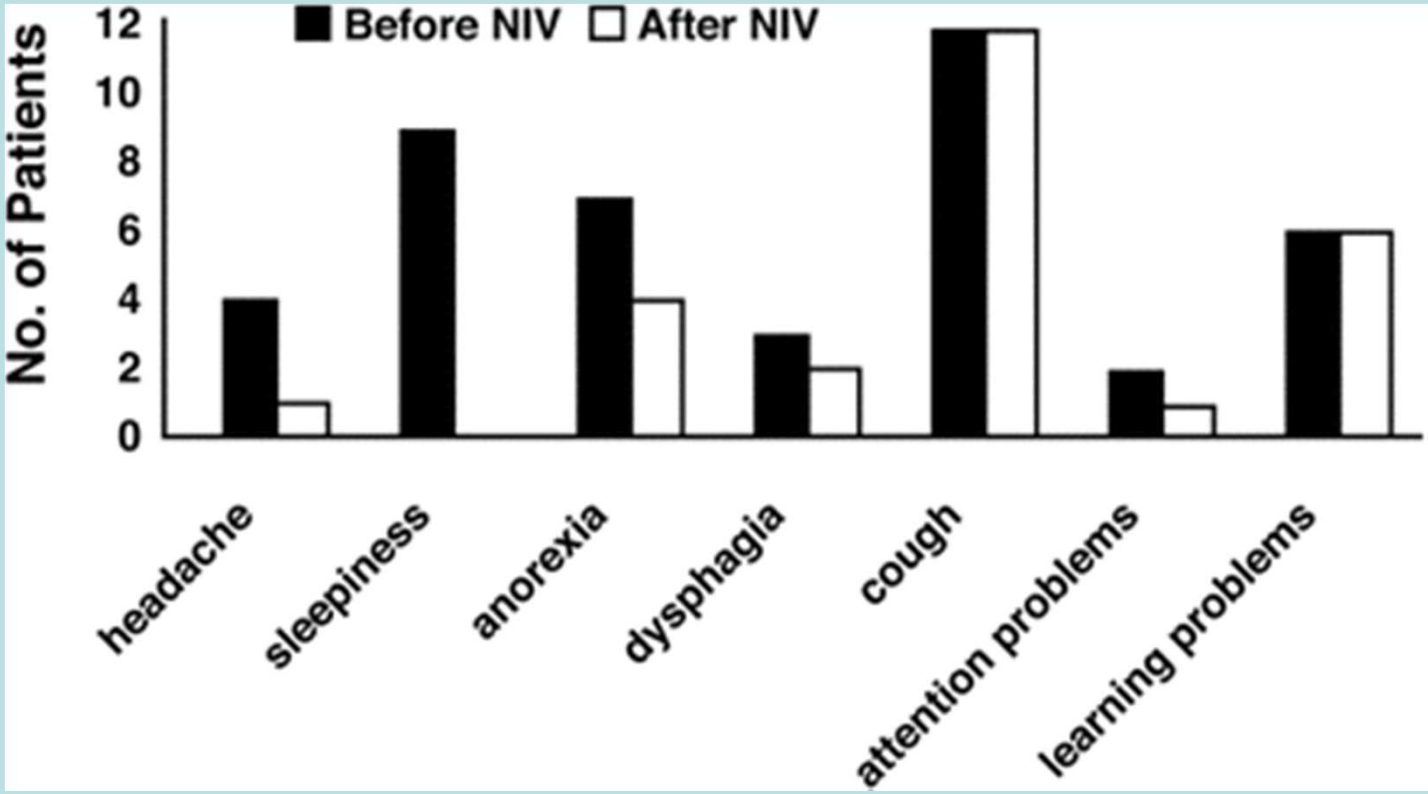


Long term NPPV is associated with an improvement in nocturnal and daytime gas exchange in children with NM disorders



Mellies *et al*, Eur Respir J 2003;22:631

Improvement in patients' symptoms after NPPV



Young *et al.* Neurology 2007;68:198

Long-term nasal intermittent positive pressure ventilation in advanced Duchenne's muscular dystrophy

A. VIANELLO, M. BEVILACQUA, V. SALVADOR, C. CARDAIOLI, E. VINCENTI

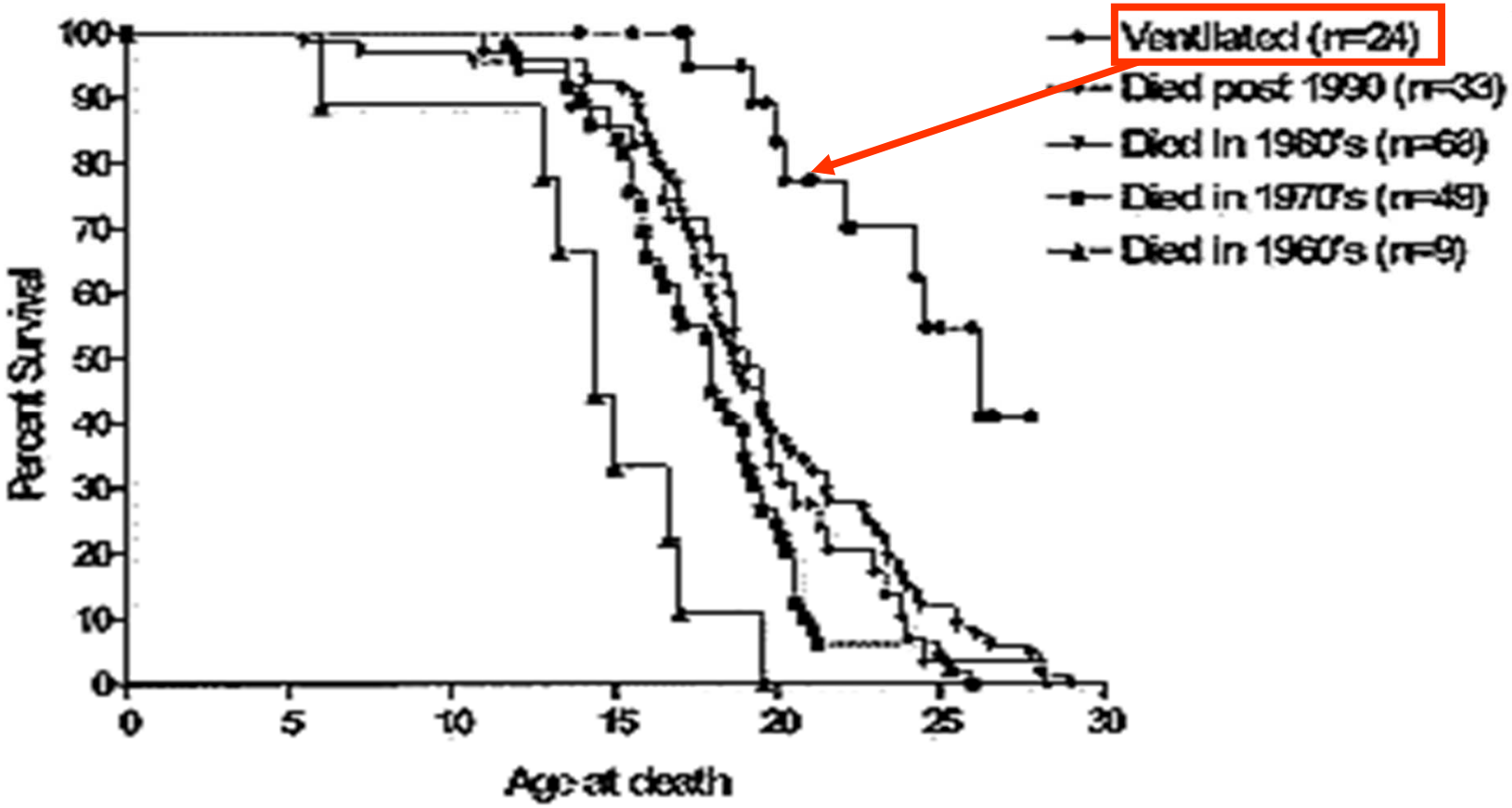
HMV IN ADVANCED DUCHENNE'S MUSCULAR DYSTROPHY

- 5 pts treated with NPPV
- 5 unventilated control pts

24 month follow-up

All pts treated with NPPV were still alive; four of five pts who underwent simple conservative treatment had died (mean survival: 9.7 ± 5.8 months)

Chest 1994;**105**:445-448



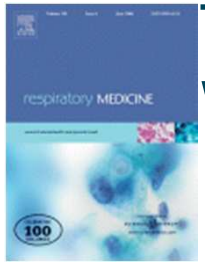
Eagle et al, Neuromusc Dis 2002

Annane D, Chevrolet JC, Chevret S, Raphael JC

Nocturnal mechanical ventilation for chronic hypoventilation in patients with neuromuscular and chest wall disorders.

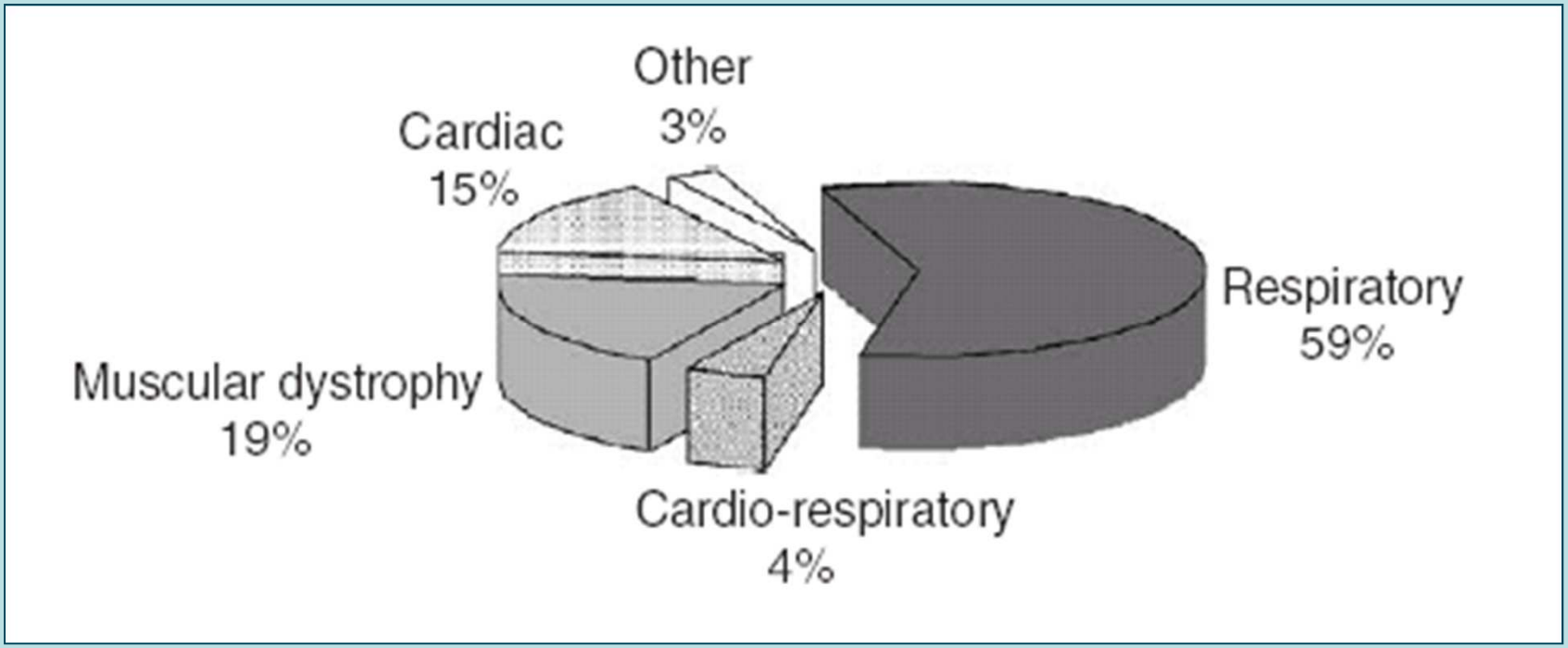
Cochrane Database of Systematic Reviews. Issue 1, 2001

Current evidence about the therapeutic benefit of mechanical ventilation is weak, but consistent, suggesting alleviation of the symptoms of chronic hypoventilation in the short term, and in two small studies survival was prolonged. Mechanical ventilation should be offered as a therapeutic option to patients with chronic hypoventilation due to neuro-muscular diseases.



Trends in survival from muscular dystrophy in England and Wales and impact on respiratory services.

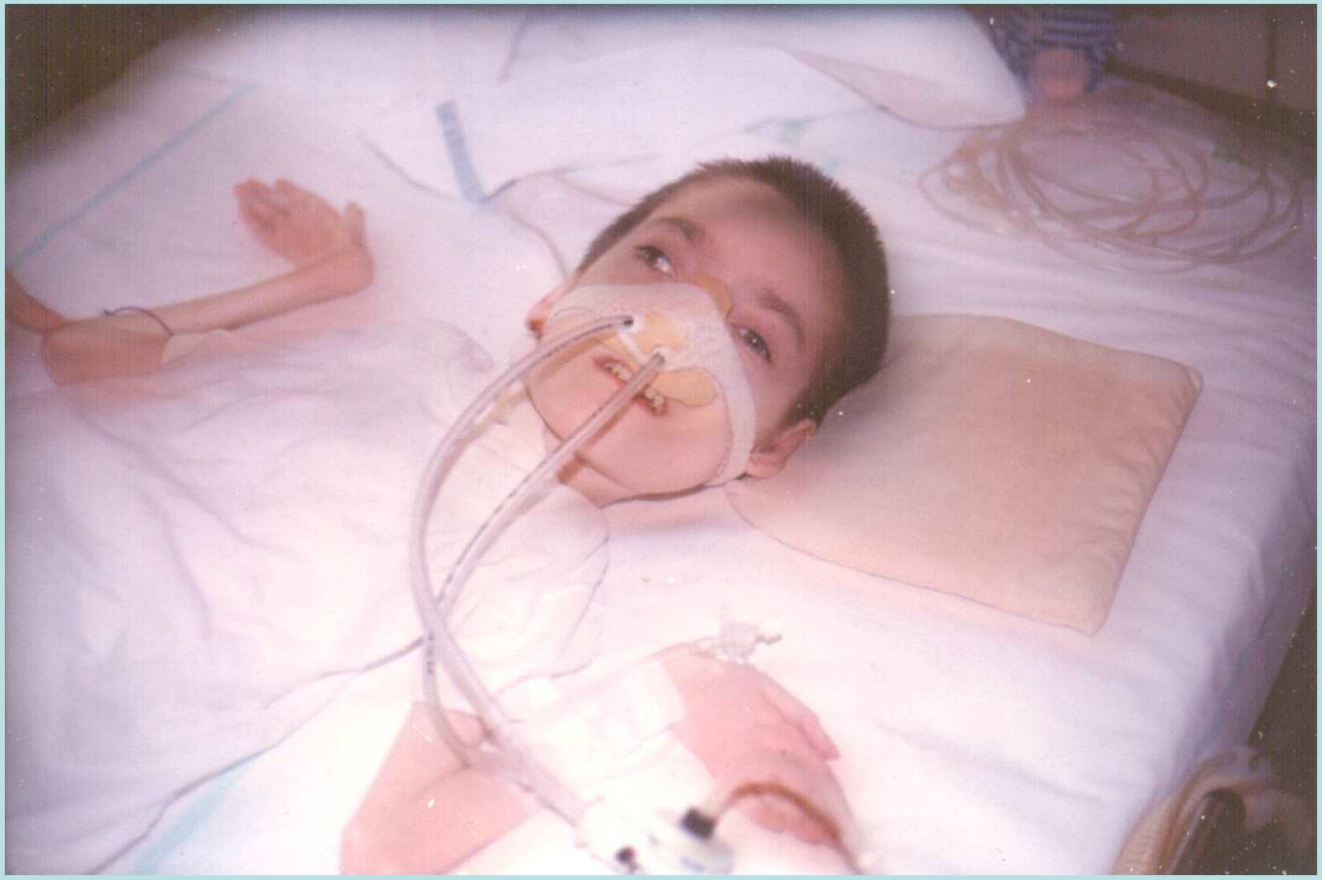
L.D. CALVERT, T.M. MC KEEVER, W.J.M. KINNEAR, J.R. BRITTON



Cause of death in muscular dystrophy in England and Wales 1993–1999

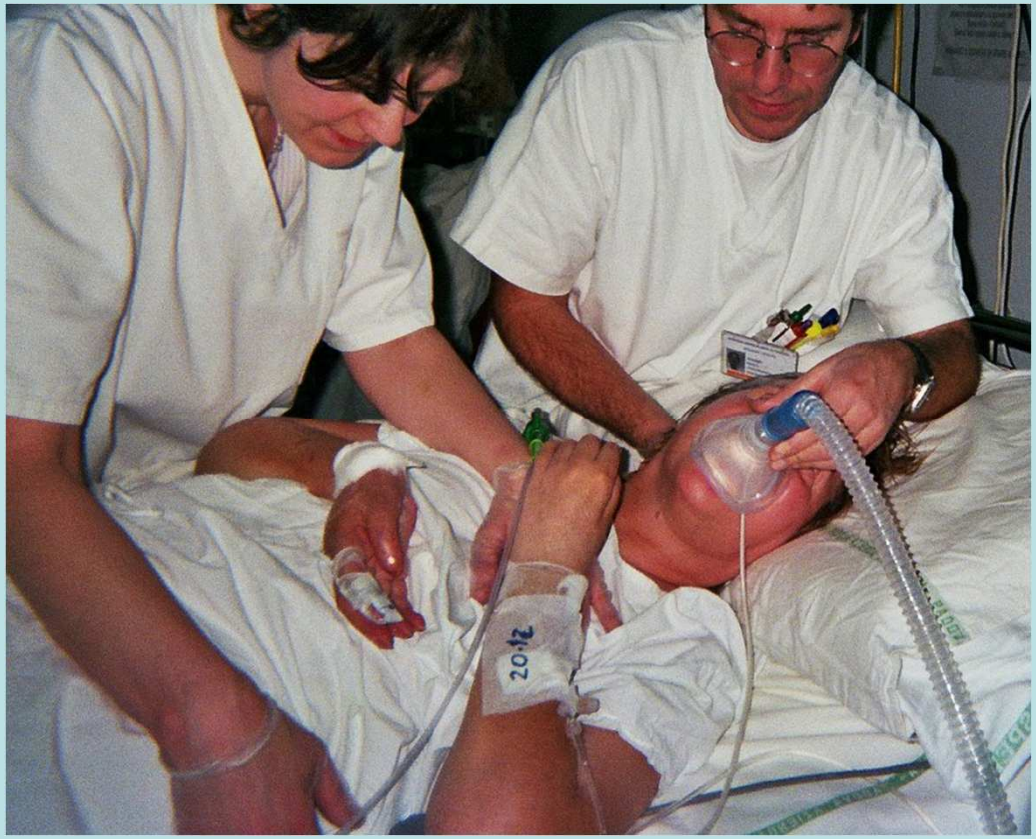
Resp Med 2005;100:1058-63

ARF in Neuromuscular patients in the Respiratory Intermediate care unit/ICU



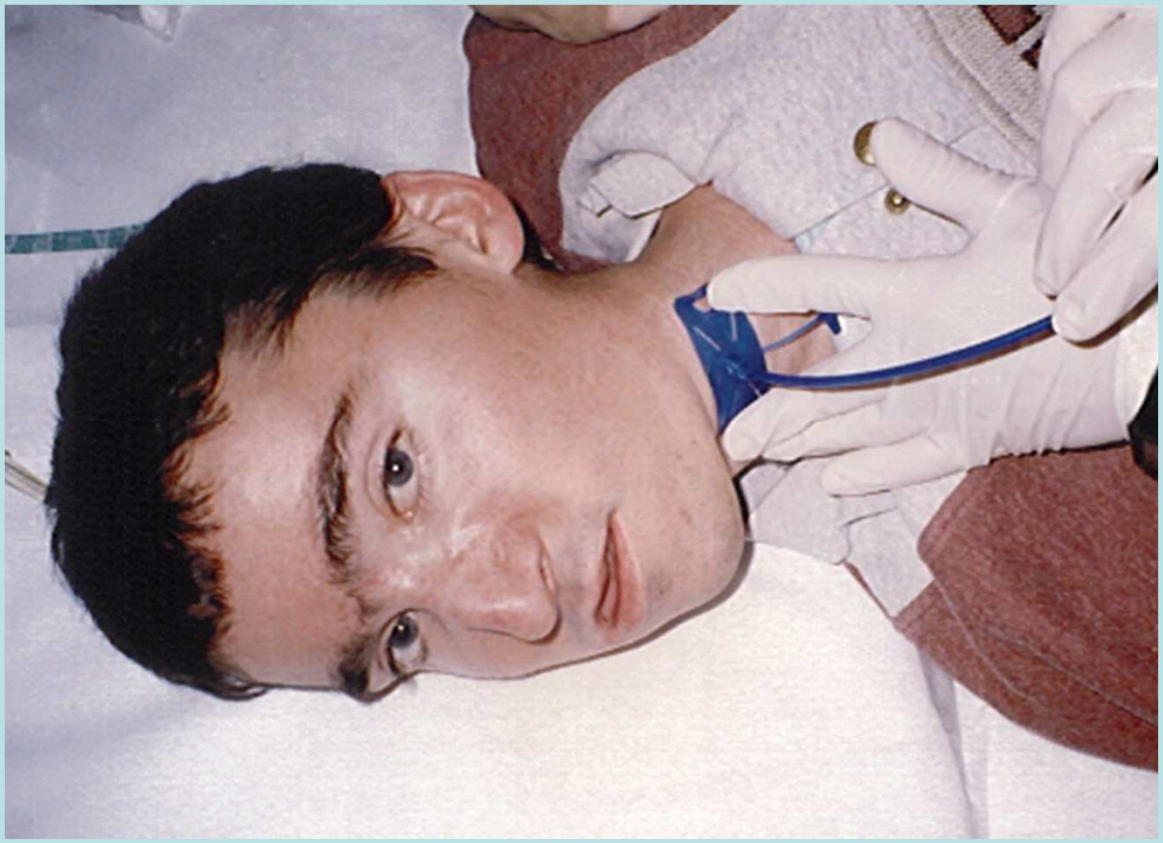
- May have extreme ventilator dependency

ARF in Neuromuscular patients in the Respiratory Intermediate care unit/ICU

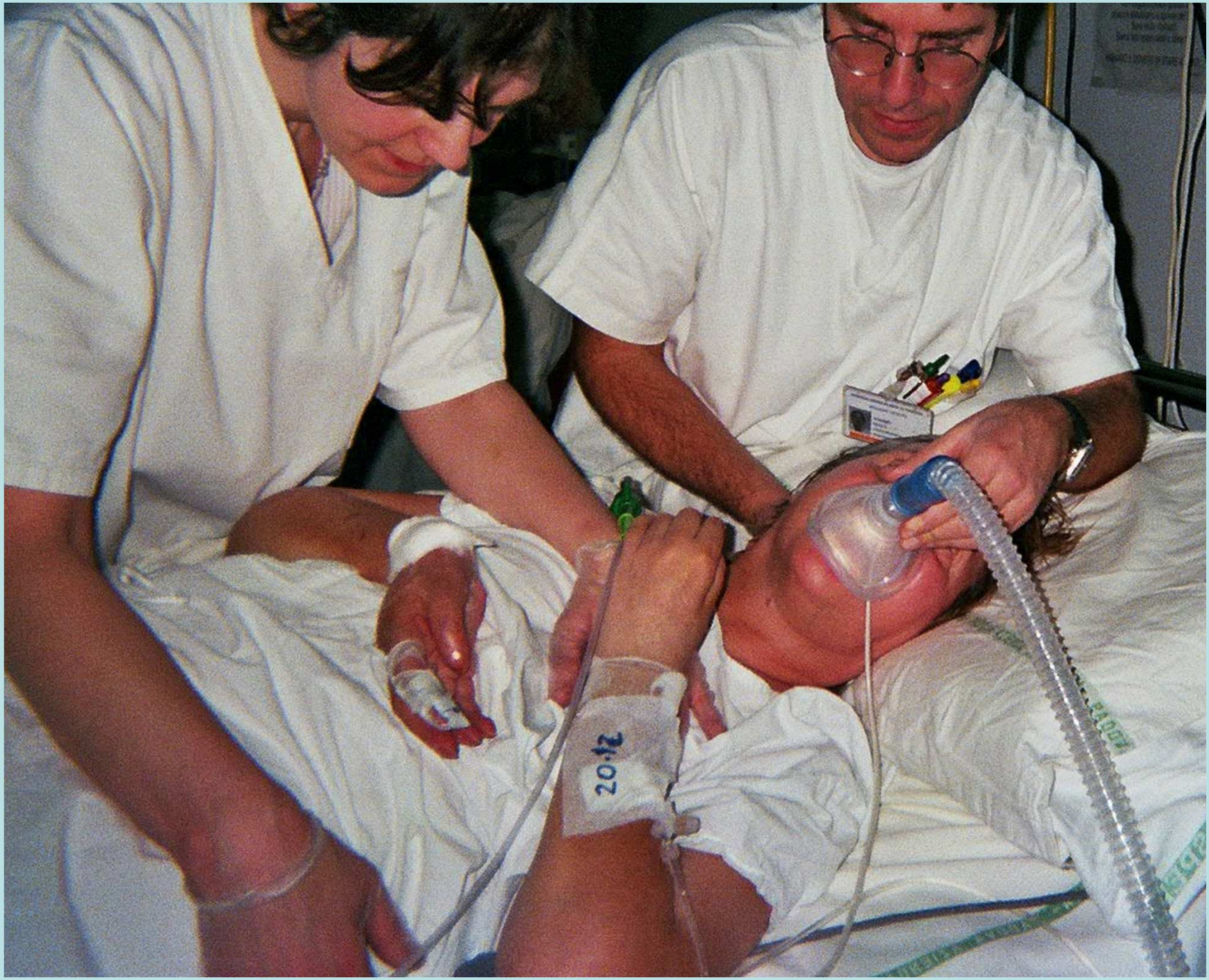


- Require intensive physiotherapy +/- cough assist

ARF in Neuromuscular patients in the Respiratory Intermediate care unit/ICU



- May have severe bulbar problems leading to intubation and tracheostomy



Major Contributors to Acute RF in Neuromuscular Disease

- Acute on chronic inspiratory muscle weakness
- Expiratory muscle weakness leading to ineffective cough
- Moderate to severe respiratory tract infection

Inspiratory muscle weakness

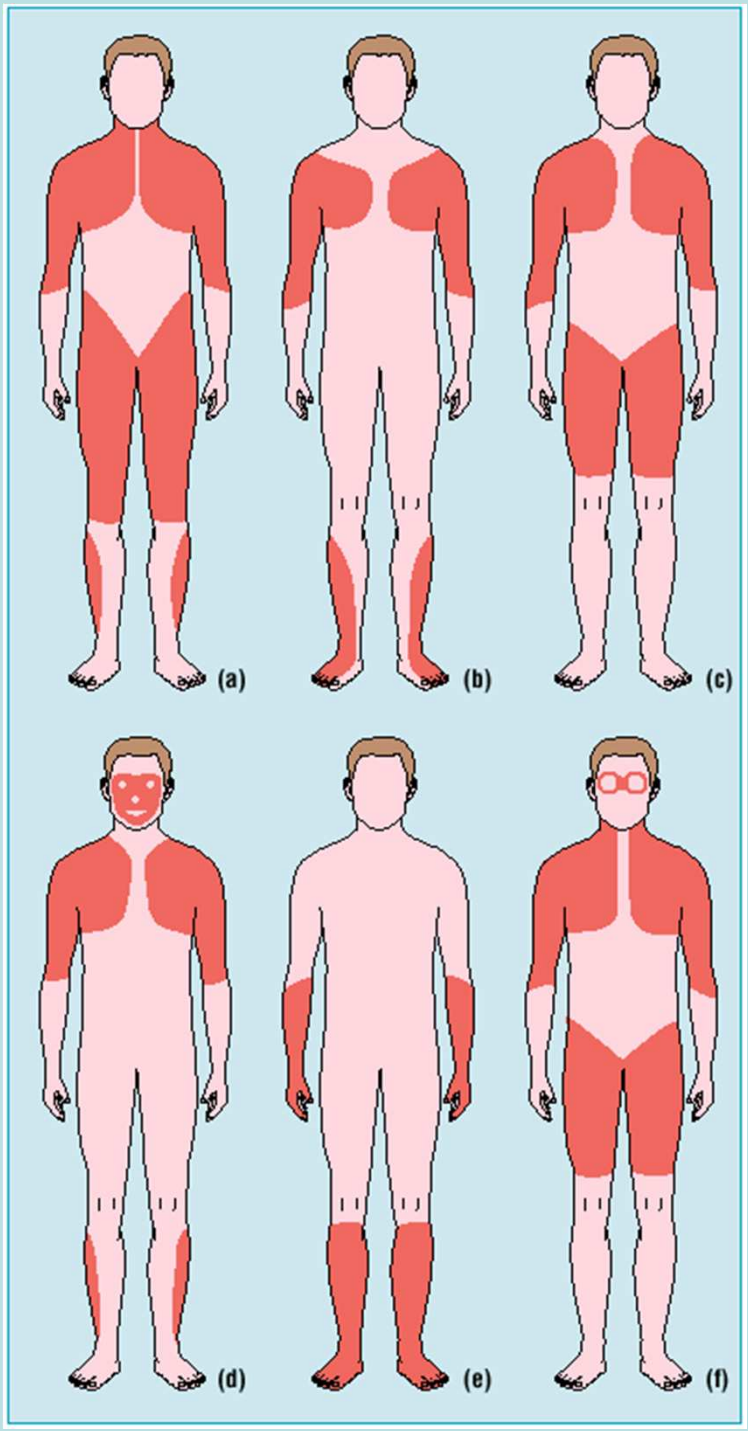
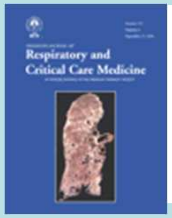


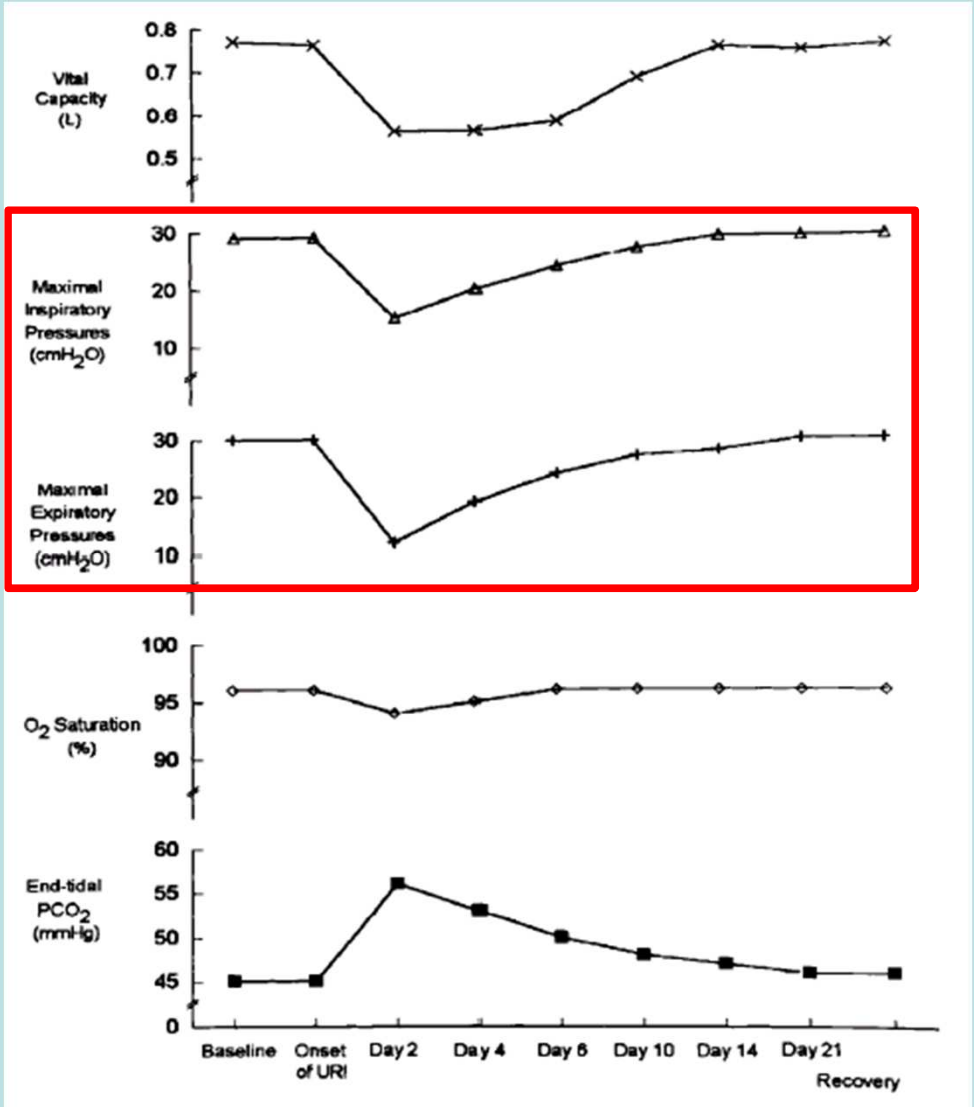
Fig 1 Distribution of predominant muscle weakness in different types of dystrophy: (a) Duchenne-type and Becker-type, (b) Emery-Dreifuss, (c) limb girdle, (d) facioscapulohumeral, (e) distal, and (f) oculopharyngeal

The muscular dystrophies
Alan E H Emery
BMJ VOLUME 317 10 OCTOBER 1998



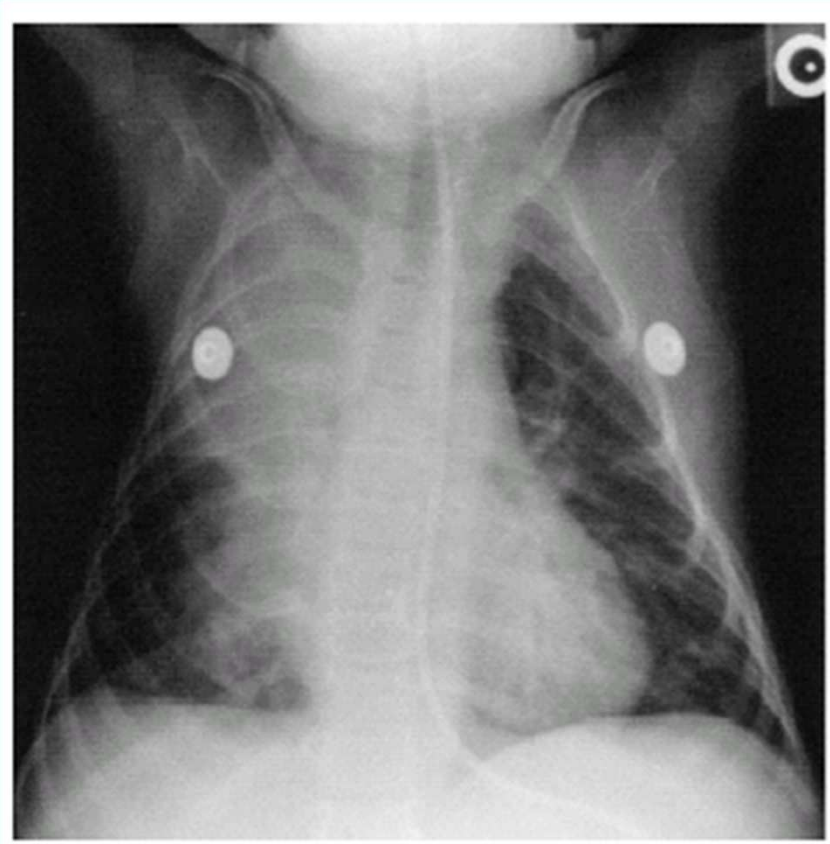
Effect of Upper Respiratory Tract Infection in Patients with Neuromuscular Disease

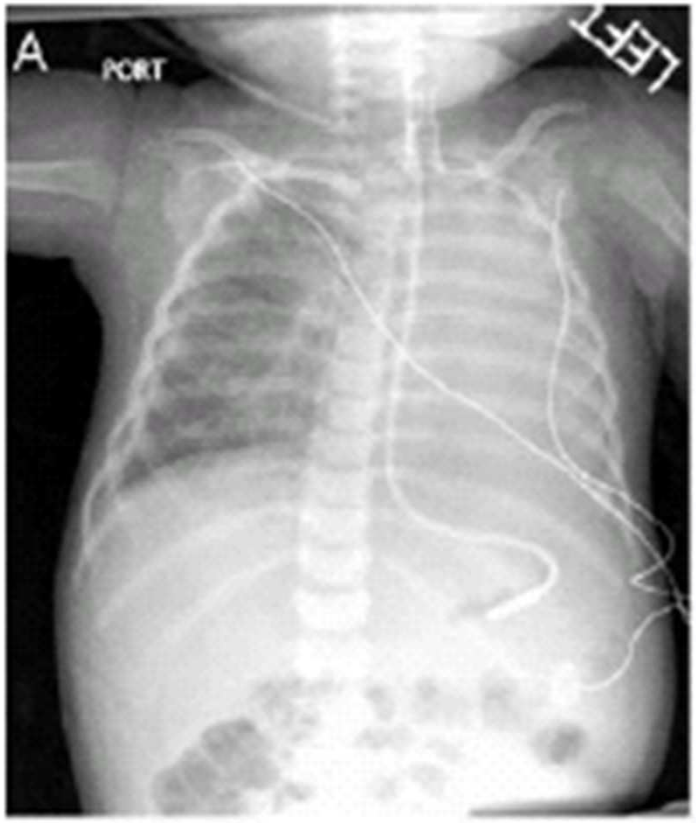
JANET M. POPONICK, I. JACOBS, GERALD SUPINSKI, and ANTHONY F. DIMARCO



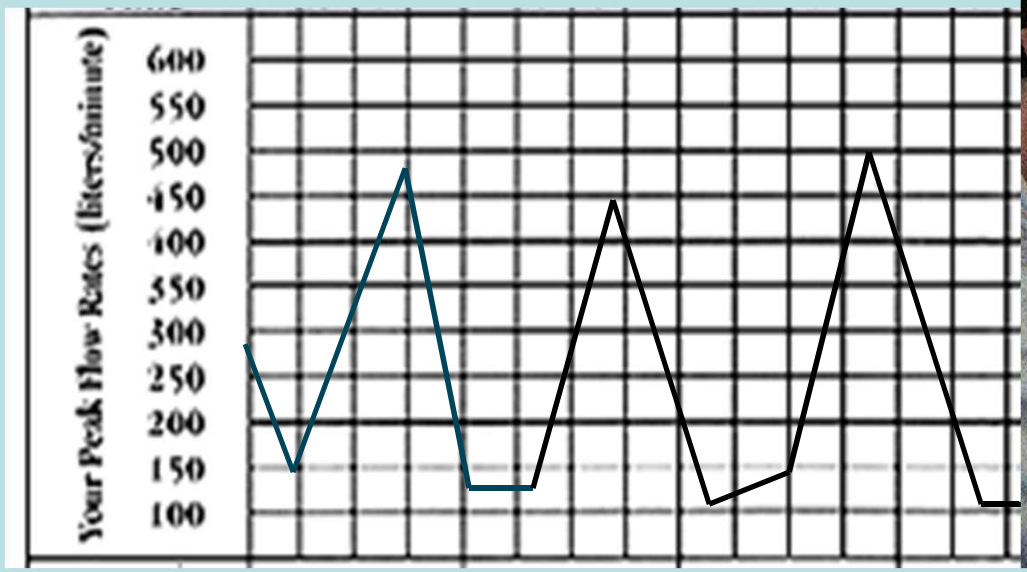
AJRCCM 1997;**156**:659-664

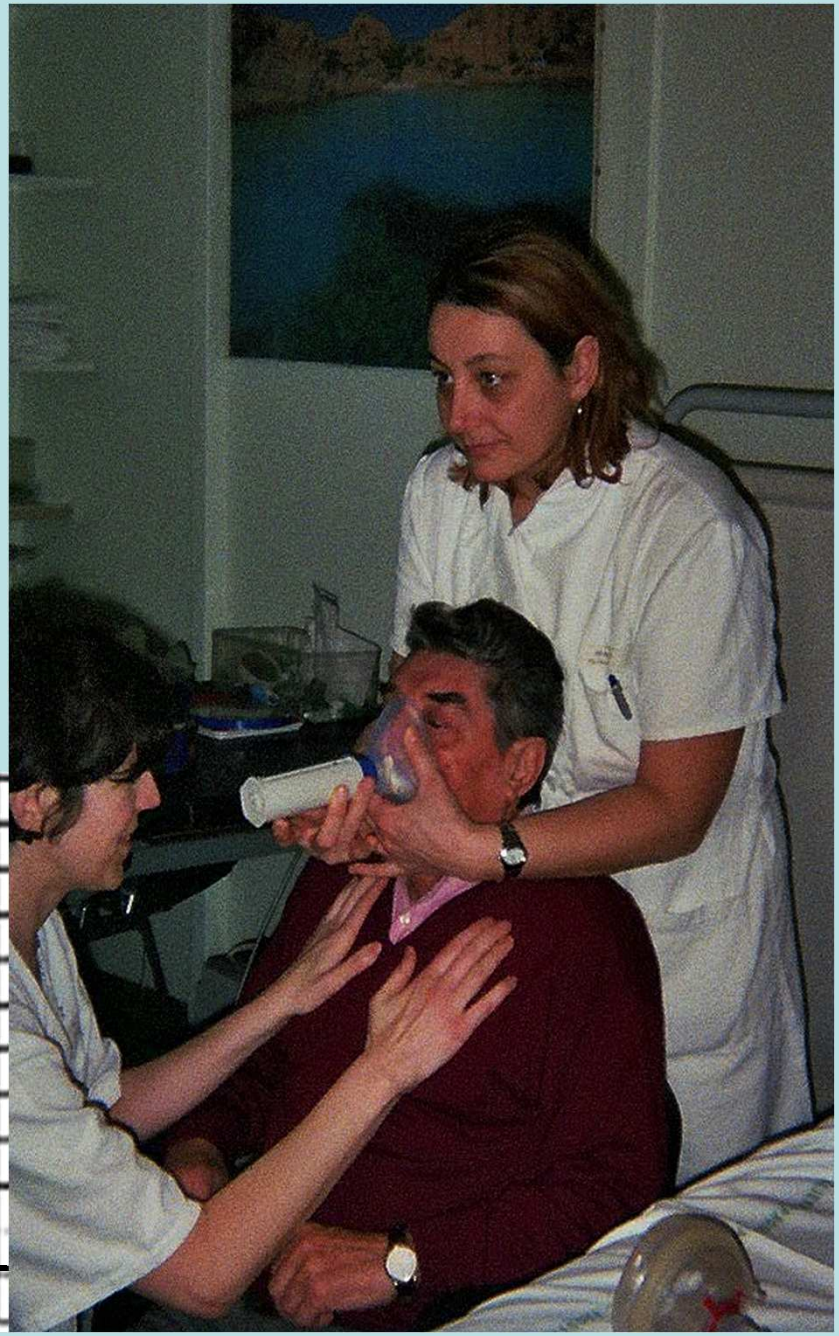
Expiratory muscle weakness leading to ineffective cough and atelectasis





Peak Cough Flow

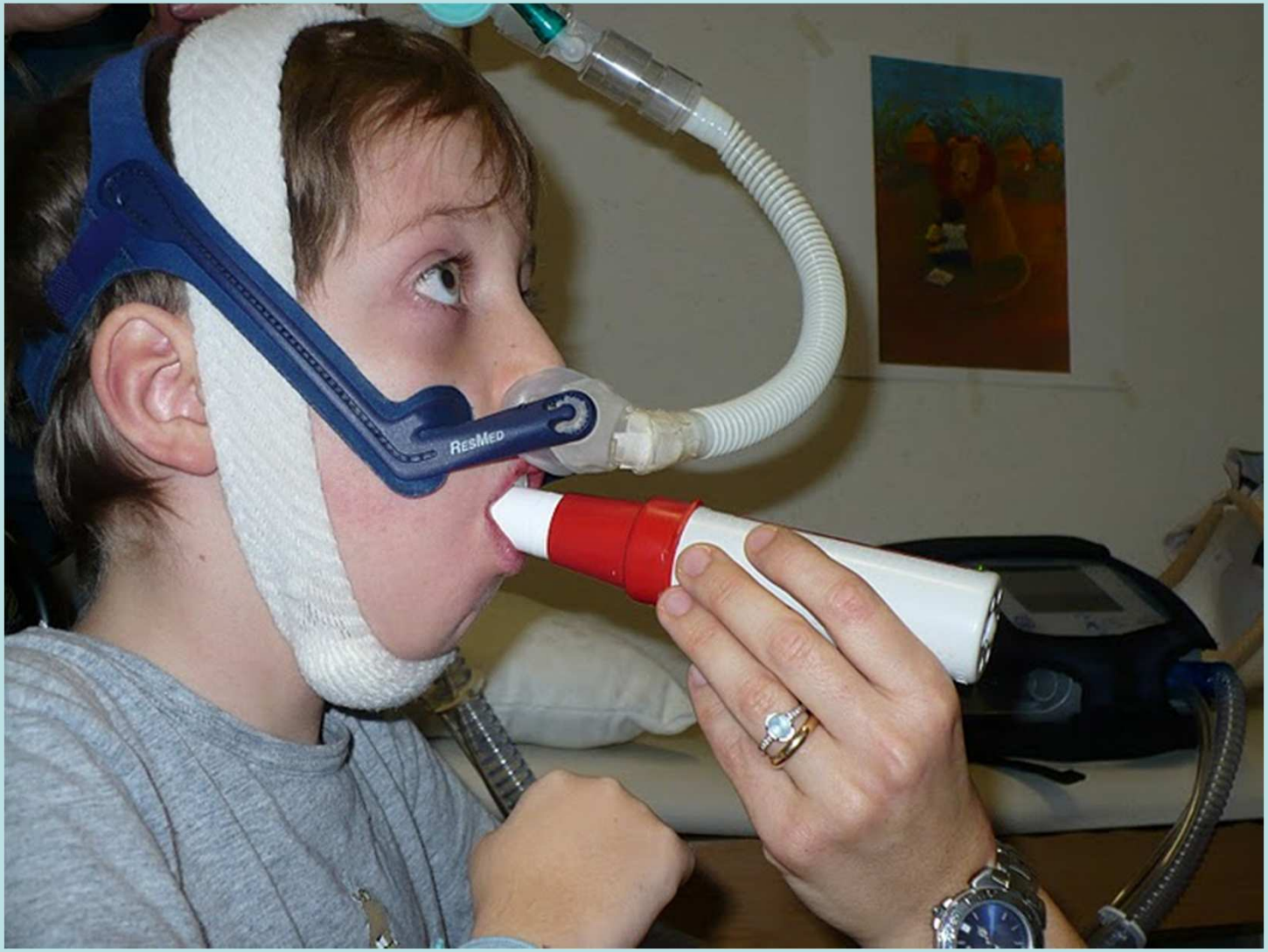




Peak Cough Flow

- Is directly correlated with the capacity to clear secretions from respiratory sistem.
- Cut-off value of 160 L/min used to identify patients who would benefit from assisted cough techniques

Bach JR , Ishikawa Y, Kim H. Prevention of pulmonary morbidity for patients with Duchenne muscular dystrophy Chest 1997;112:1024-8

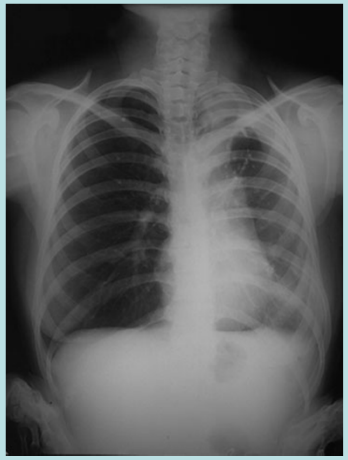

ResMed

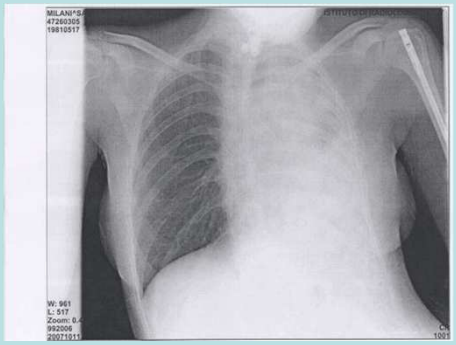
ELASTIC LOAD

↓ LUNG VOLUME

↓ LUNG COMPLIANCE

PNEUMONIA, ATELECTASIS



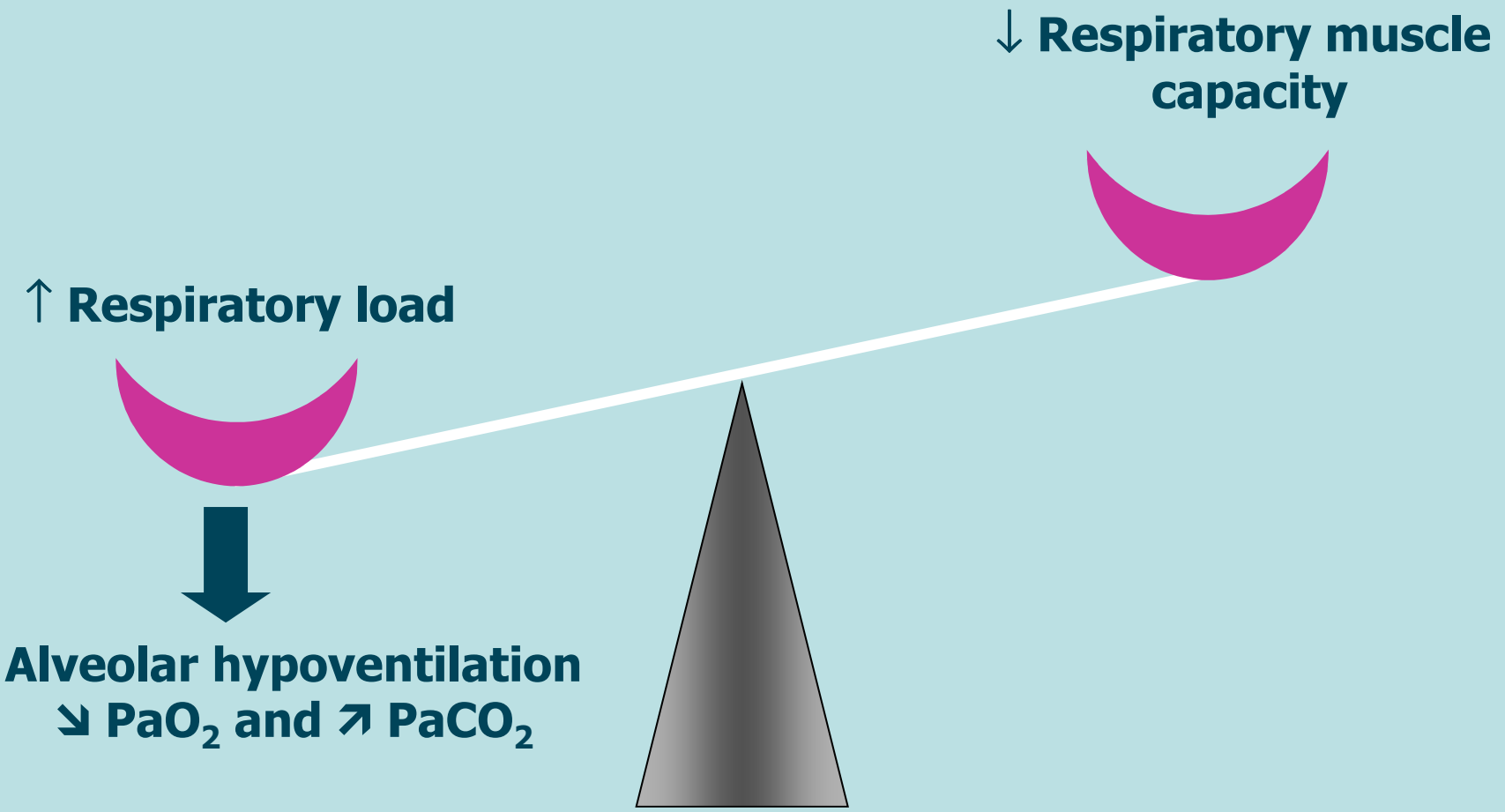


RESISTIVE LOAD

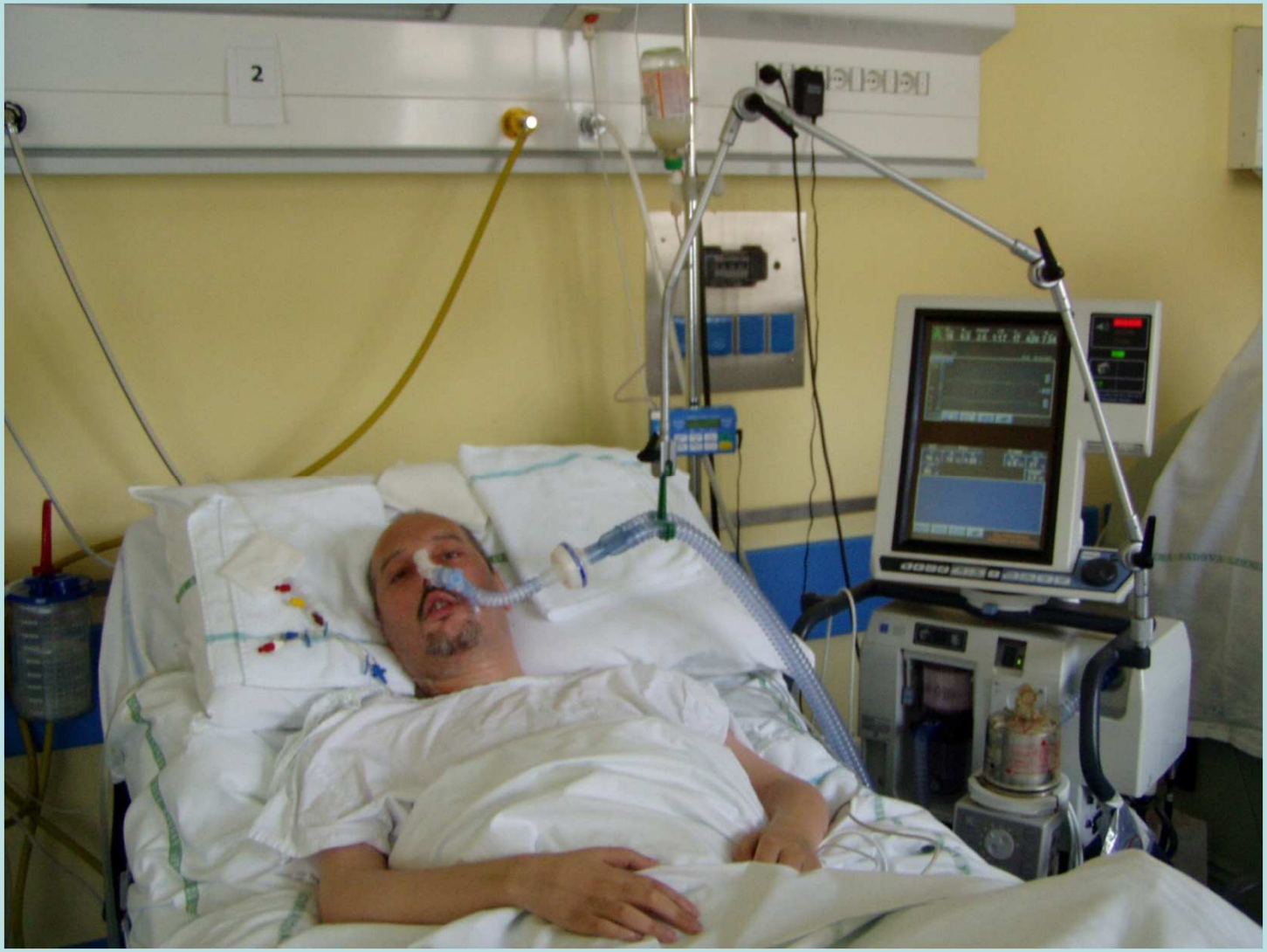
MUCOUS ENCUMBRANCE

↑ AIRWAY RESISTANCE

PHYSIOPATHOLOGY OF ACUTE RESPIRATORY FAILURE



Conventional Approach to Managing ARF in Neuromuscular Disorders

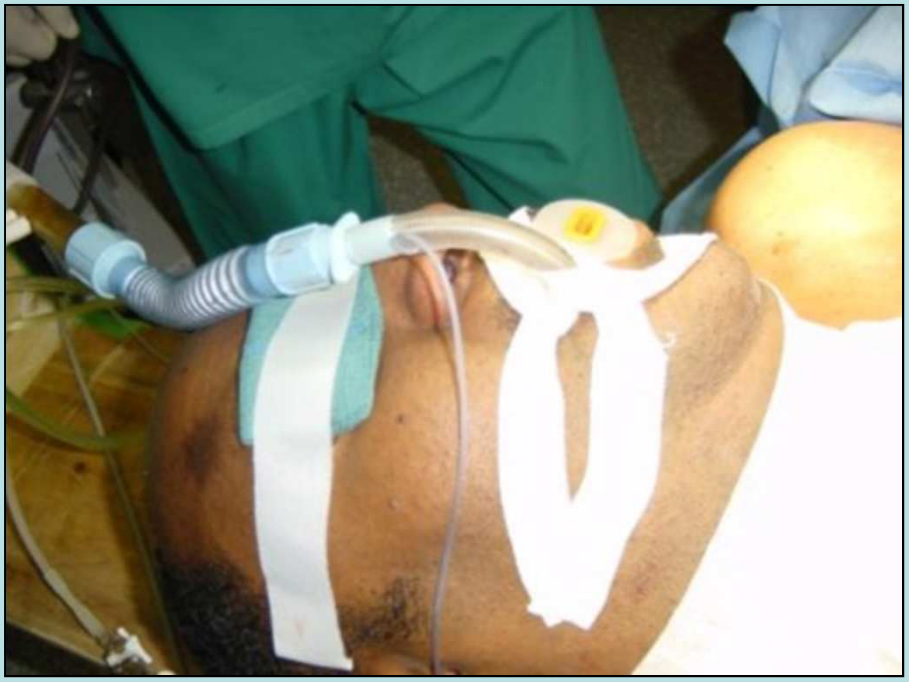


Complications associated with intubation and invasive mechanical ventilation

Cardiac arrest
Generalized seizures
Gastric distension
Mechanical dysfunction of the endotracheal tube
Cuff leaks
Self-extubation
Respiratory muscle dysfunction
Respiratory muscle atrophy
Decrease in cardiac output
Barotrauma
Increase in work of breathing
Nosocomial infections (sinusitis, VAP)
Injury to the pharynx, larynx, and trachea (ulceration, oedema, haemorrage, stenosis, loss of voice)
Weaning difficulties

Management of Tracheal Intubation in the Respiratory Intensive Care Unit by Pulmonary Physicians

Andrea MA Vianello MD, Giovanna ME Arcaro MD, Fausto S Braccioni MD, Federico Gallan MD, Chiara M Greggio MD, Anna Marangoni MD, Carlo Ori MD, and Michele Minuzzo MD



In 60 cases intubations were performed successfully.
Complications occurred in 4 cases:

all were patients with neuromuscular disorders.

Respiratory Care • January 2007 Vol 52 No 1

Outcome of ventilatory support for acute respiratory failure in motor neurone disease

M D Bradley, R W Orrell, J Clarke, A C Davidson, A J Williams, D M Kullmann, N Hirsch, R S Howard

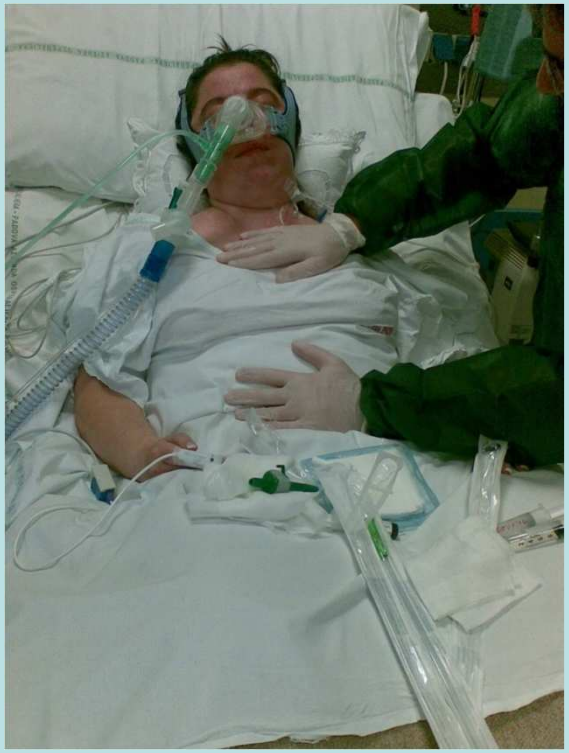
Table 2 Outcomes of 24 patients with MND at the end of the study period

	Previously diagnosed (n=7)	Not previously diagnosed (n=17)	Total (n=24)
Died on ITU while receiving ventilatory support	2 (29%)	5 (29%)	7 (29%)
Time (range) spent on ITU before death (days)	25–56	7–54	7–56
Discharged from ITU with respiratory support	5 (71%)	12 (71%)	17 (71%)
Fully extubated and discharged with no support	0	1 (6%)	1 (4%)
IPPV through tracheostomy	4 (57%)	9 (53%)	13 (54%)
IPPV through facemask	1 (14%)	1 (6%)	2 (8%)
Rocking bed	0	1 (6%)	1 (4%)

IPPV, intermittent positive pressure ventilation; ITU, intensive therapy unit

J Neurol Neurosurg Psychiatry 2002;**72**:752–756

CRITICAL ISSUE RAISED BY THE CONVENTIONAL MANAGEMENT OF ARF IN NMD



Should NIV be considered a safer and more effective alternative to endotracheal intubation as a first-line intervention?

A. Vianello
M. Bevilacqua
G. Arcaro
F. Gallan
E. Serra

Intensive Care Med (2000) 26: 384–390

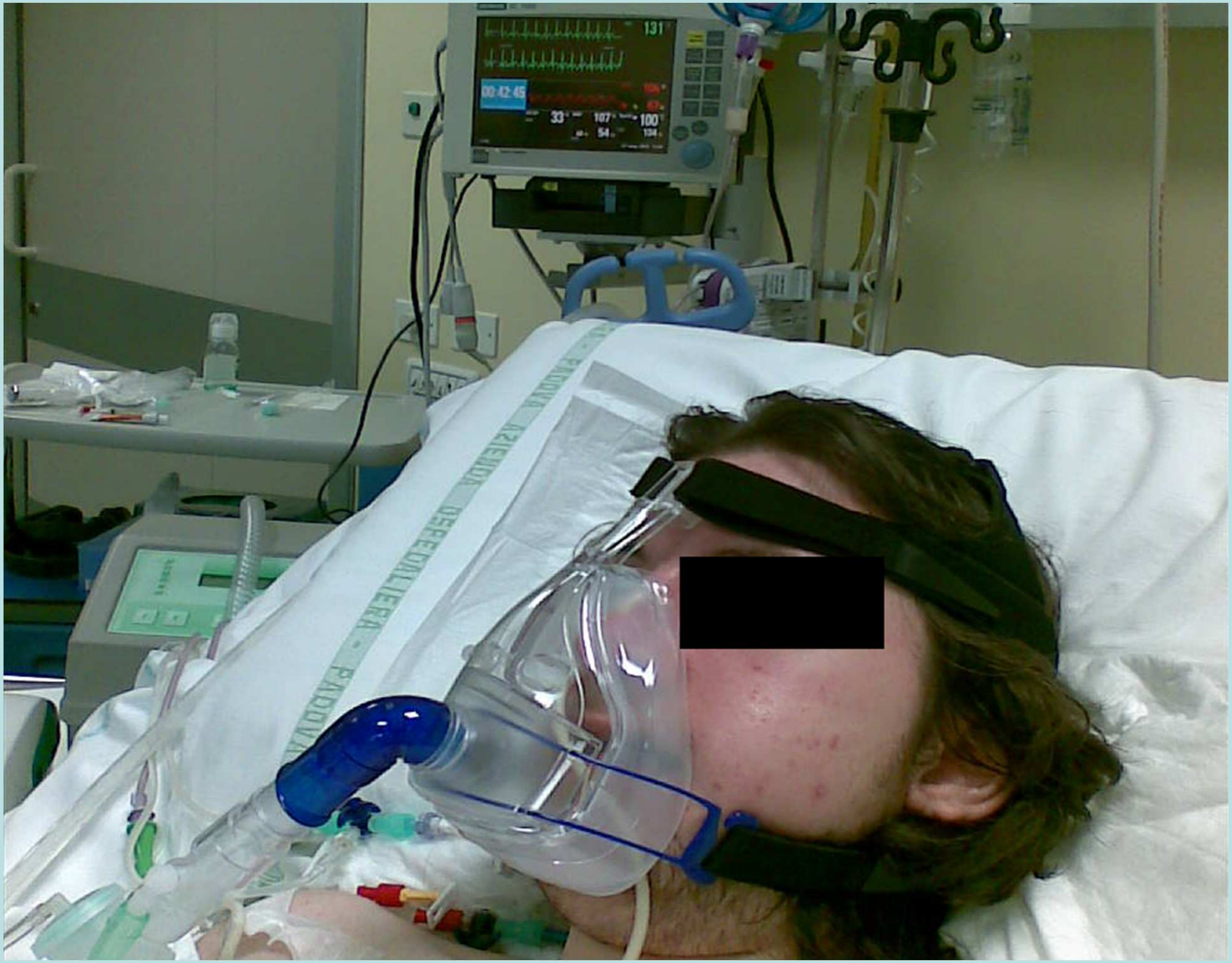
Non-invasive ventilatory approach to treatment of acute respiratory failure in neuromuscular disorders. A comparison with endotracheal intubation

- Period of study: from 1995 to 1998
- Type of study: controlled; historically matched control patients
- Patient populations: 14 consecutive patients with Neuromuscular Disorders in ARF in whom MV was mandatory
- All patients were treated with NPPV as first line of treatment
- Intubation or tracheostomy was provided when NPPV failed

Intensive Care Med 2000;**26(4)**:384-390

Neuromuscular Disorders

Type	No of Patients
• MD Duchenne	7
• ALS	4
• Congenital Myopathy	1
• Congenital Muscular Dystrophy	1
• Motor-Sensory Neuropathy	1
Total	14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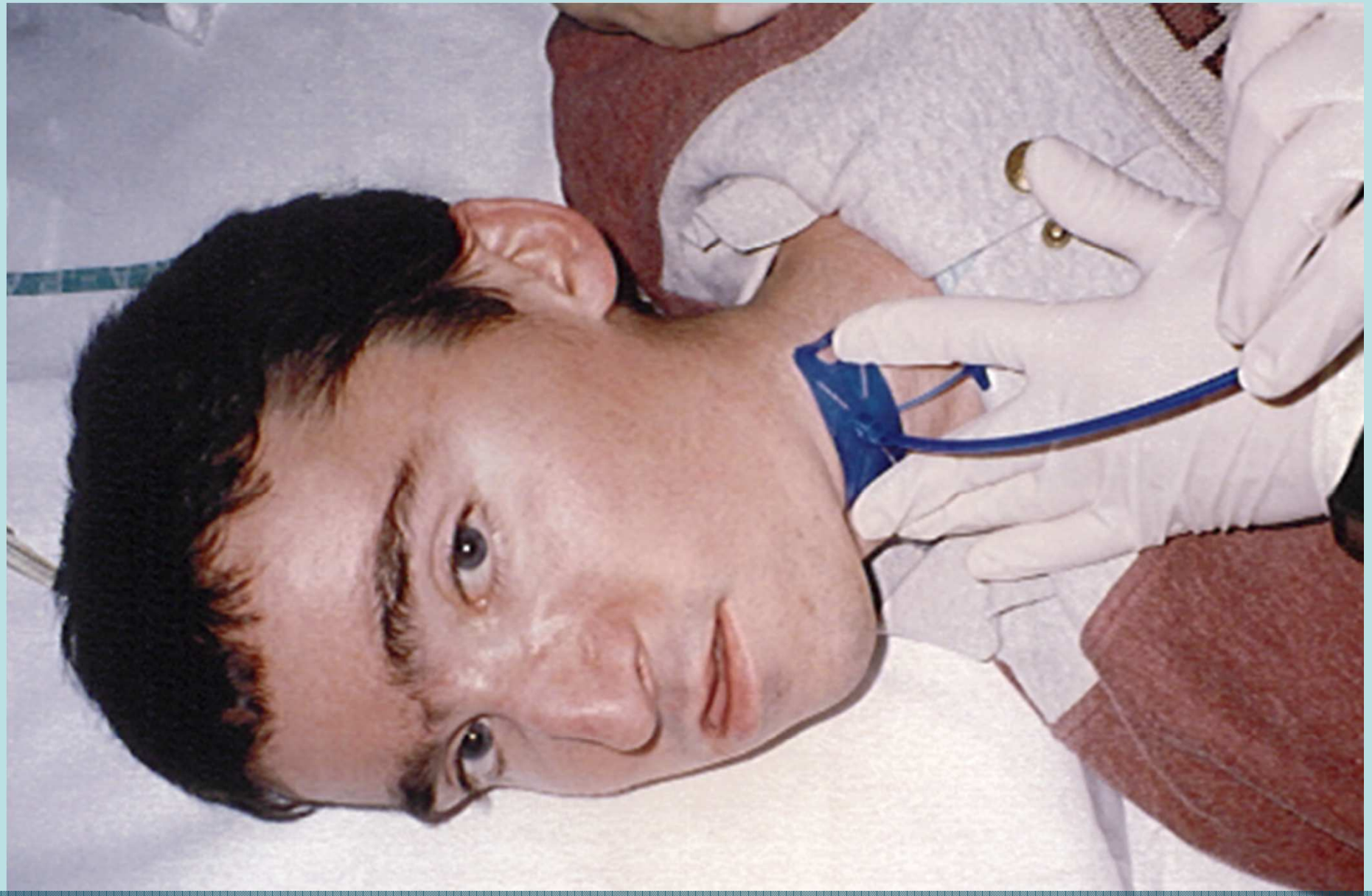
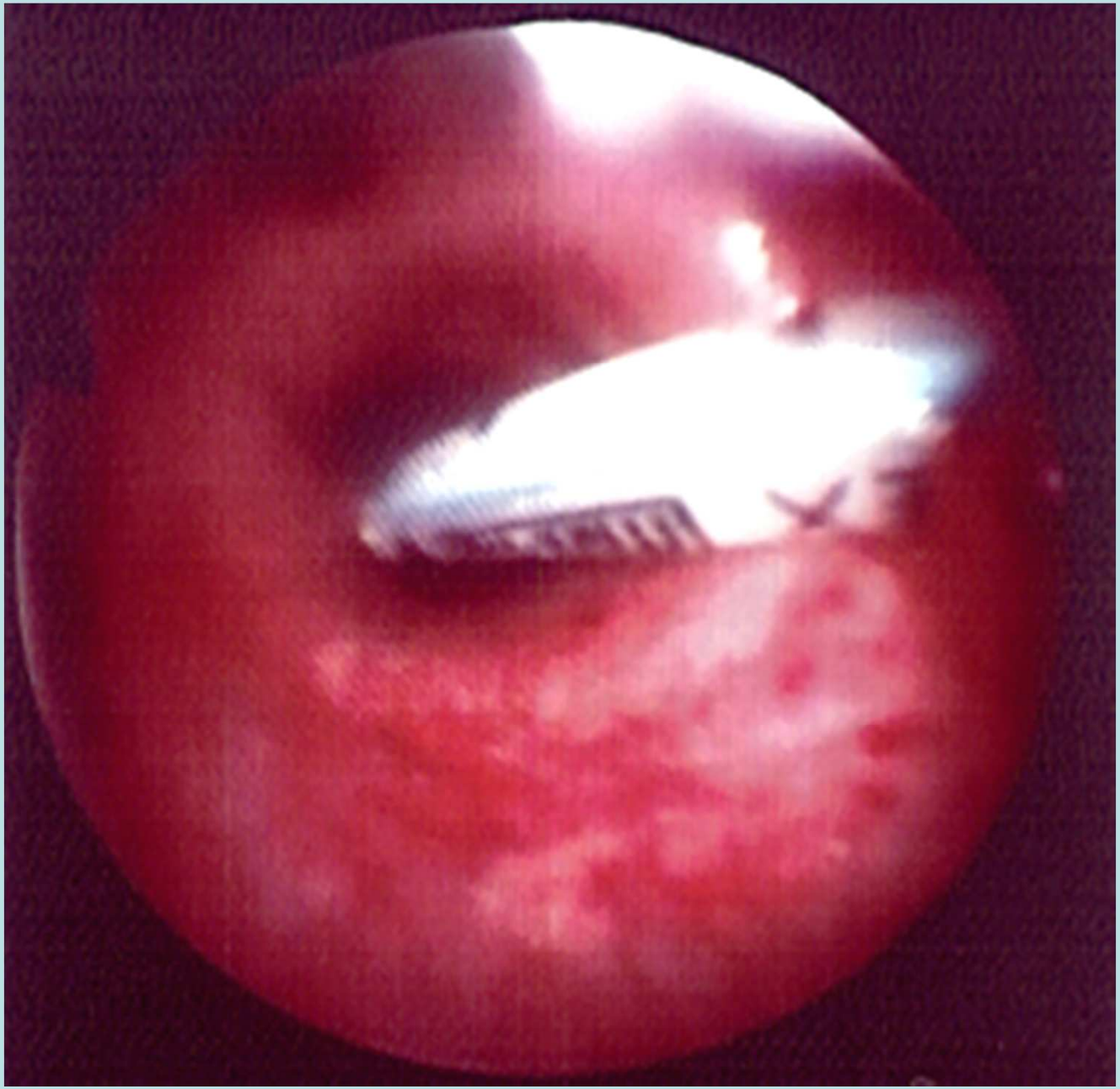


CRICOTHYROID MINITRACHEOSTOMY



Study Endpoints

1. Mortality
2. Treatment Failure
3. Time to improvement
4. Length of ICU Stay

Clinical Outcome of Patients: Cumulative Data

	Group A	Group B	P Value
Death, No	2	8	0.046
Treat. Failure, No	4	11	0.021
Time to improvement, hrs	8.4 (2.8)	2.8 (11)	0.0001
ICU stay, days	14.9 (10.7)	47.1(51.9)	0.032

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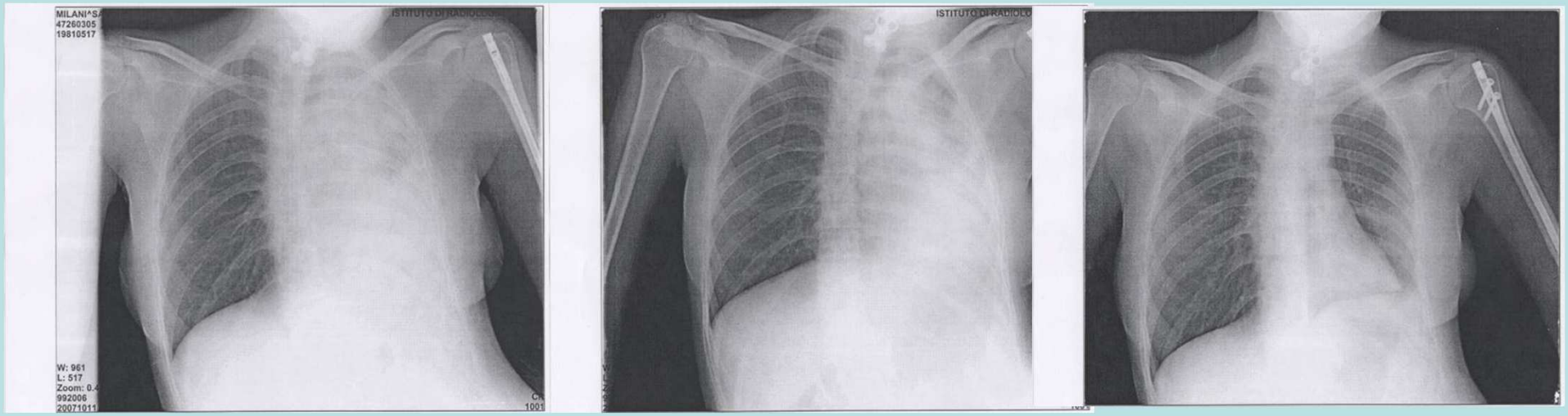
Intensive Care Med (2000) 26: 384–390

Non-invasive ventilatory approach to treatment of acute respiratory failure in neuromuscular disorders. A comparison with endotracheal intubation

CONCLUSIONS

- The application of NIV tends to reduce mortality and treatment failure in comparison with PPV via ETI
- The use of NIV combined with cricothyroid "minitracheostomy" in neuromuscular ARF could be extended to patients with ineffective cough
- However, the ability to adequately protect the upper airway is crucial to the success and patient selection remains important

Cough assist: mechanical in-exsufflation (M-IE)



NIV assisted physio, ambu bag, portable suction machine

Addition of cough inexsufflator:

→ Cough PF <160l/min, poor cough

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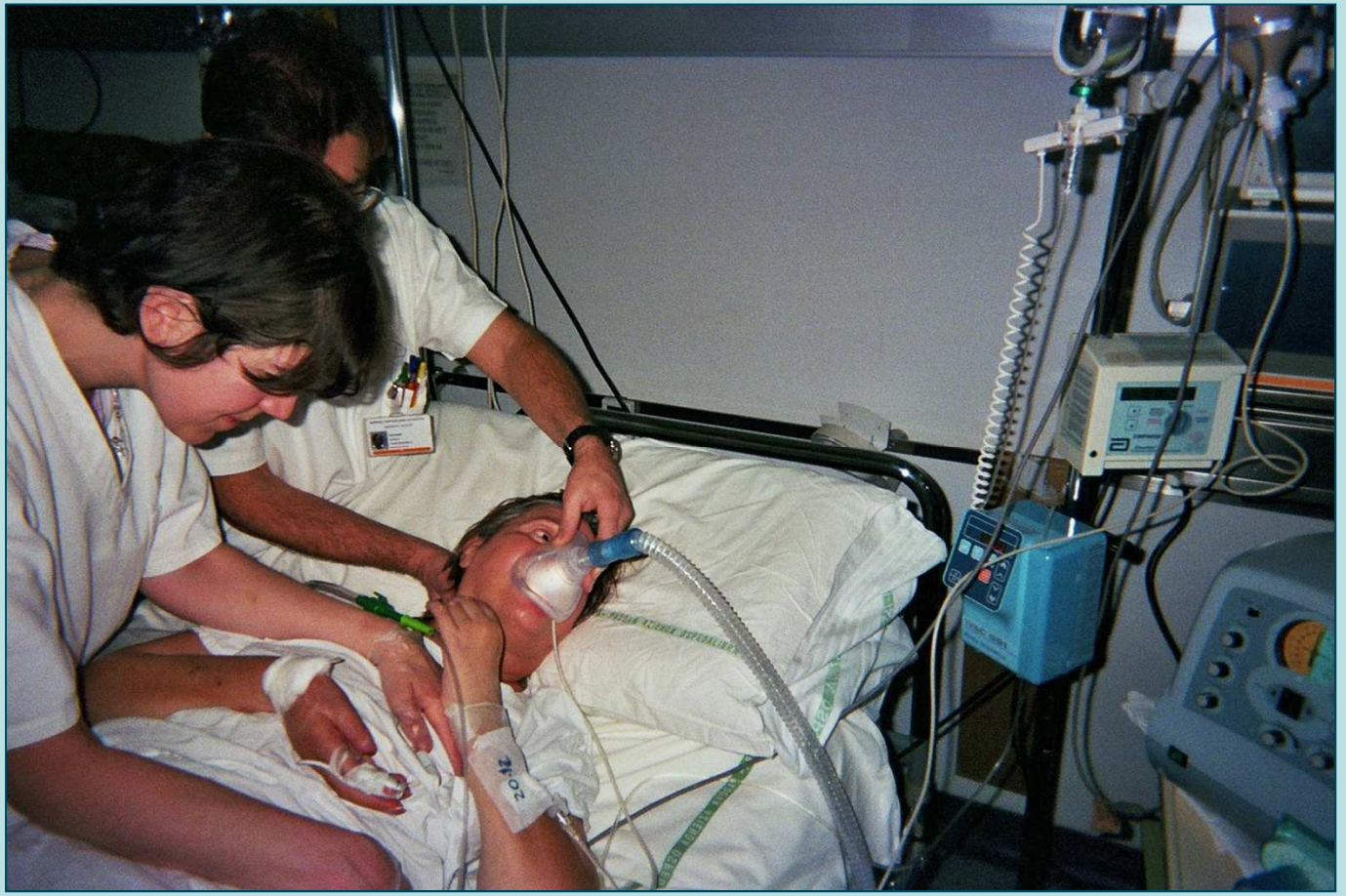
Am J Phys Med Rehabil, 2005;**84**:83-88

Mechanical Insufflation–Exsufflation Improves Outcomes for Neuromuscular Disease Patients with Respiratory Tract Infections



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Am J Phys Med Rehabil, 2005;**84**:83-88

Mechanical Insufflation–Exsufflation Improves Outcomes for Neuromuscular Disease Patients with Respiratory Tract Infections

- Period of study: from January 2001 to March 2003
- Type of study: controlled
- Patient populations: 11 consecutive neuromuscular patients with URTI and mucous encumbrance
- All patients were treated with MI-E in addition to conventional CPT
- Cricothyroid "mini-tracheostomy" or endotracheal intubation was considered when MI-E plus CPT could not expulse airway secretions

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Am J Phys Med Rehabil, 2005;**84**:83-88

Mechanical Insufflation–Exsufflation Improves Outcomes for Neuromuscular Disease Patients with Respiratory Tract Infections

	Group A	Group B	p Value
Time spent on MV (days)	9.4 ± 6.9	13.5±11.9	n.s.
Hospital stay (days)	20.5 ±20	19.8 ± 17	n.s.
Treatment failure, N.	2	10	< 0.05
Pts who required BAA, N.	5	6	n.s.

Group A: Mechanical In-Exsufflator + Chest Physical Treatment
Group B: Chest Physical Treatment

Miguel R. Goncalves, BS
John R. Bach MD

Mechanical Insufflation–Exsufflation Improves Outcomes for Neuromuscular Disease Patients with Respiratory Tract Infections

A Step in the Right Direction

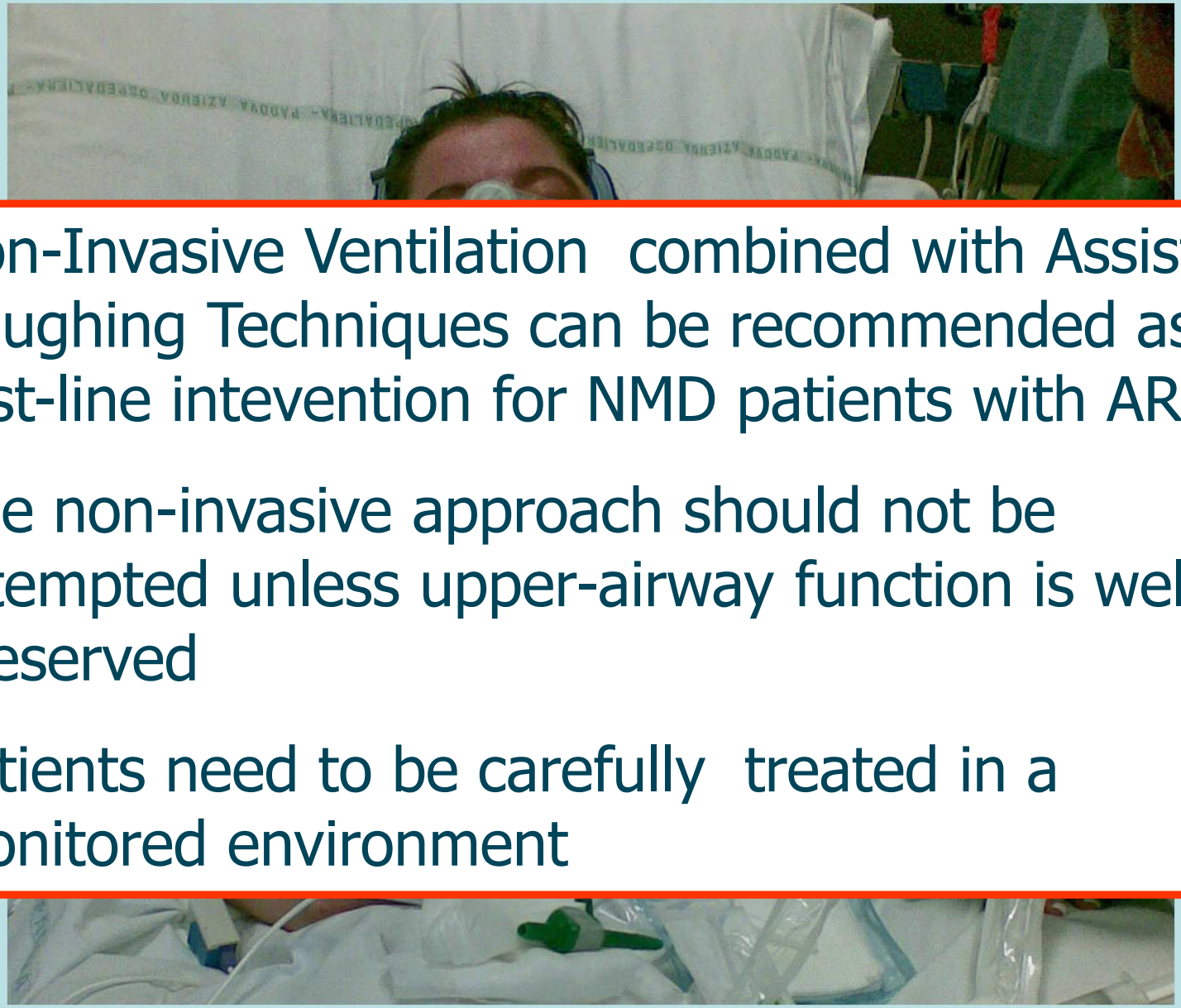
Am J Phys Med Rehabil, 2005;**84**:89-91

COMMENT

If both the inspiratory and expiratory muscle aids are used effectively, only advanced bulbar ALS and some SMA type 1 patients who develop ARF require intubation and tracheostomy.

CRITICAL ISSUE RAISED BY THE CONVENTIONAL MANAGEMENT OF ARF IN NMD

Should non-invasive ventilatory approach be considered a safer and more effective alternative to endotracheal intubation and invasive ventilation?



1. Non-Invasive Ventilation combined with Assisted Coughing Techniques can be recommended as a first-line intevention for NMD patients with ARF
2. The non-invasive approach should not be attempted unless upper-airway function is well preserved
3. Patients need to be carefully treated in a monitored environment

Conclusions

LT NIV is extremely effective in prolonging survival and improving quality of life in NMD patients with CRF

Acute Respiratory Failure actually represents the leading cause of death in patients with NMD

Non-Invasive management of respiratory complication is possible and effective even in the acute setting