

Intervenció terapèutica precoç: com, quan, té sentit?



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Abordatje respiratori precoç en el malalt amb ELA



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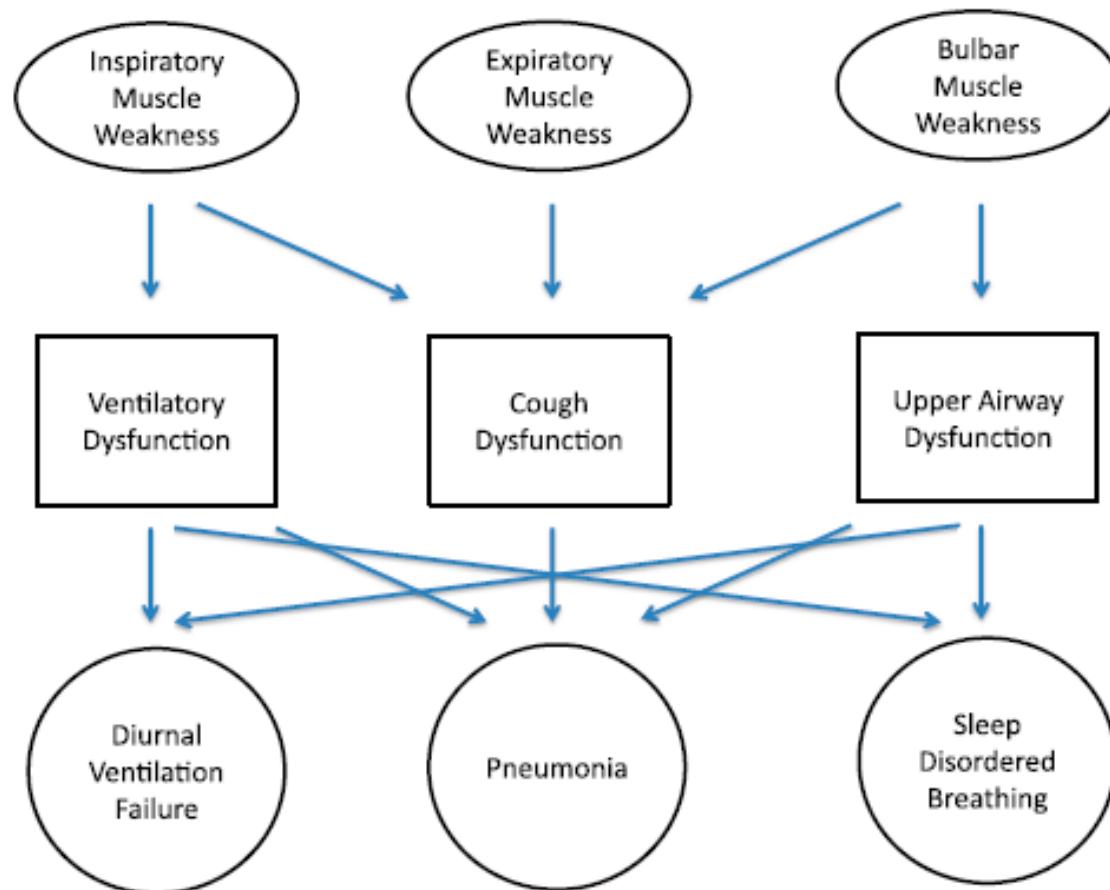
Concise Clinical Review

Pulmonary Issues in Patients with Chronic Neuromuscular Disease

Joshua O. Benditt¹ and Louis J. Boitano¹

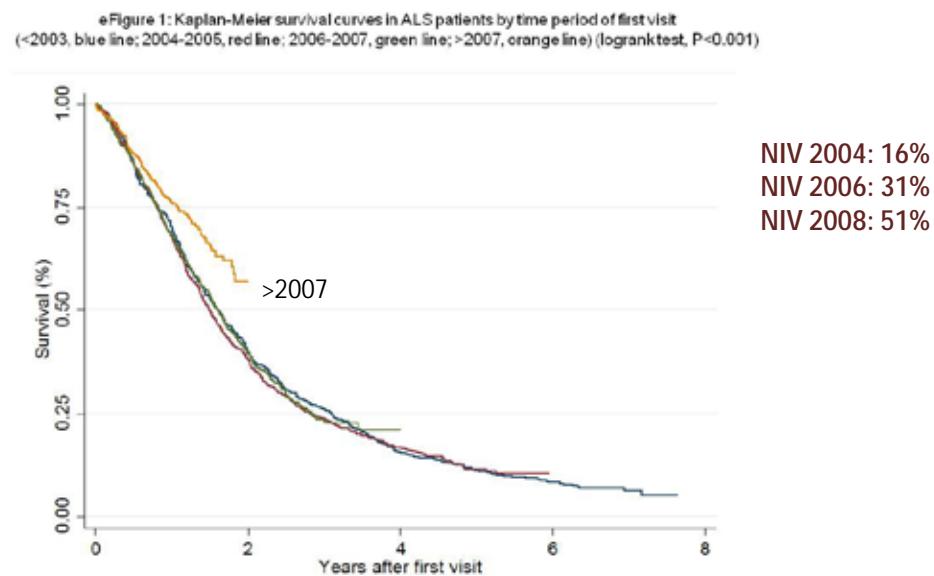
¹University of Washington School of Medicine, Seattle, Washington

Am J Respir Crit Care Med Vol 187, Iss. 10, pp 1046–1055, May 15, 2013



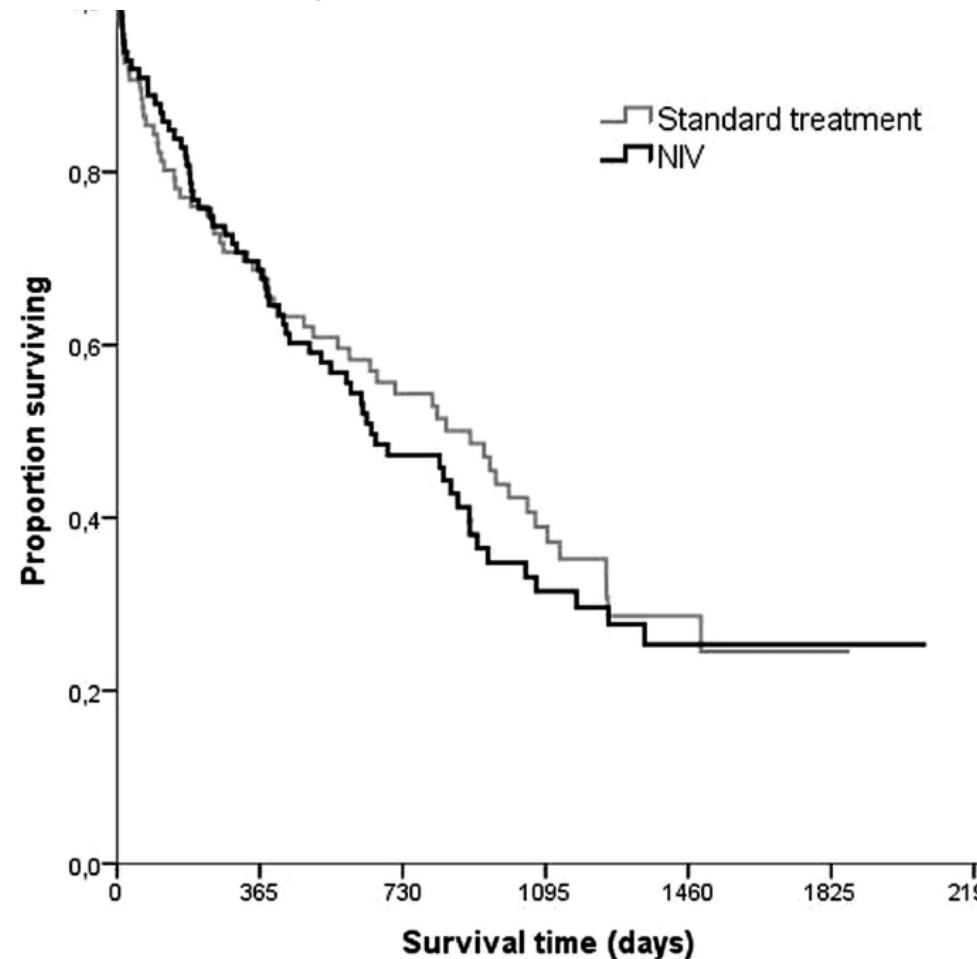
JUSTIFICACIÓ de la intervenció respiratoria

- La ELA no es cura , PERO ES TRACTA
(Dr Diaz Lobato, *dixit*)



Nocturnal non-invasive ventilation in COPD patients with prolonged hypercapnia after ventilatory support for acute respiratory failure: a randomised, controlled, parallel-group study

F M Struik,^{1,2} R T M Sprooten,³ H A M Kerstjens,^{1,2} G Bladder,¹ M Zijnen,⁴ J Asin,⁵ N A M Cobben,³ J M Vonk,^{2,6} P J Wijkstra^{1,2}



Abordatje diagnòstic precoç

Abordatge diagnòstic antic

- Visita a demanda (si clínica)
- Visió “funcionalista”
- Manca de comunicació entre professionals
- Manca de coneixement dels desitjos del malalt i la família
- Dificultat en la presa de decisions
- Manca de temps

Normativa SEPAR

Normativa sobre el manejo de las complicaciones respiratorias de los pacientes con enfermedad neuromuscular

Guidelines for the Management of Respiratory Complications in Patients With Neuromuscular Disease

Eva Farrero^a, Antonio Antón^b, Carlos J. Egea^c, M. José Almaraz^d, J. Fernando Masa^e, Isabel Utrabo^e, Miriam Calle^f, Héctor Verea^g, Emilio Servera^h, Luis Jaraⁱ, Emilia Barrotⁱ y Vinyet Casolivé^a

^a Unidad Funcional Interdisciplinaria Sociosanitaria (UFISS) Respiratoria, Hospital de Bellvitge, L'Hospitalet de Llobregat, Barcelona, España

^b Servicio de Respiratorio, Hospital de la Santa Creu i Sant Pau, Barcelona, España

^c Unidad Sueño y Ventilación, S. Respiratorio, Hospital Universitario de Álava, Álava, España

^d Equipo de Soporte de Cuidados Paliativos, Hospital Universitario de Álava, Álava, España

^e Servicio de Neumología, Hospital San Pedro de Alcántara, Cáceres. CIBERES, ISCIII, Madrid, España

^f Servicio de Respiratorio, Hospital de San Carlos, Madrid, España

^g Complejo Hospitalario Universitario A Coruña, CHUAC, A Coruña, España

^h Unidad de Cuidados Respiratorios, Servicio de Neumología, Hospital Clínico Universitario de Valencia, Valencia, España

ⁱ Unidad Médico-Quirúrgica de Enfermedades Respiratorias, Hospital Universitario Virgen del Rocío, Sevilla, España

VALORACIÓN INICIAL

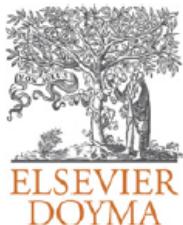
1. Respiratoria: valoración clínica, técnicas de imagen (Rx simple, radioscopía, otros), función pulmonar (gases arteriales, espirometría, pico flujo de la tos, PIM, PEM, SNIF, pruebas en decúbito (opcional), volúmenes (opcional), difusión (opcional), estudio nocturno (plusioximetría, PLSG).
2. Estudio de disfagia: valoración clínica, pruebas de cribaje, videofluoroscopia.
3. Valoración de complicaciones cardiovasculares (si procede).



SEGUIMIENTO

1. Enfermedades neuromusculares de lenta evolución:
 - seguimiento cada 6-12 meses.
 - evaluación síntomas, espirometría, GSA, SNIF/PIM/PEM, pulsioximetría.
2. Enfermedades neuromusculares de rápida evolución:
 - seguimiento cada 2-4 meses.
 - evaluación síntomas, espirometría, pico flujo de la tos, gases arteriales, SNIF, pulsioximetría (opcional).
 - seguimiento en consulta multidisciplinar (recomendable).

COMITÉ ELA



ARCHIVOS DE BRONCONEUMOLOGÍA

www.archbronconeumol.org



Artículo especial

Atención integral a pacientes con esclerosis lateral amiotrófica: un modelo asistencial

Maria Rosa Güell^{a,*}, Antonio Antón^a, Ricardo Rojas-García^b, Carmen Puy^a
y Jesus Pradas^b, en representación de todo el grupo interdisciplinario

^a <org>Departamento de Neumología</org>, <addL>Hospital de la Santa Creu i Sant Pau</addL>, Barcelona, España

^b <org>Unidad de Enfermedades Neuromusculares</org>, <addL>Servicio de Neurología, Hospital de la Santa Creu i Sant Pau</addL>, Barcelona, España



SANT PAU

11

COMITÈ E.L.A.





7







EVOLUCIÓ GASOS EN SANG ARTERIAL

EDAT (anys):

PES Pak

ALÇADA (cm):

DIA	9.6-06				
O ₂ (%)	21%				
pH	7.414				
PaO ₂ (mmHg)	82				
PaCO ₂ (mmHg)	46				
Sat Hb (%)	96.6				
Sat Hb-O ₂ (%)	95.2				
Hb (g./100 ml)	14.2				
CO.Hb (%)	0.9				
HCO ₃ st (mmol/L)	27.8				
HCO ₃ real (mmol/L)	28.6				
Excess Base (mmol/L)	4.3				
DIF(A-a)PO ₂ (mmHg)	16				

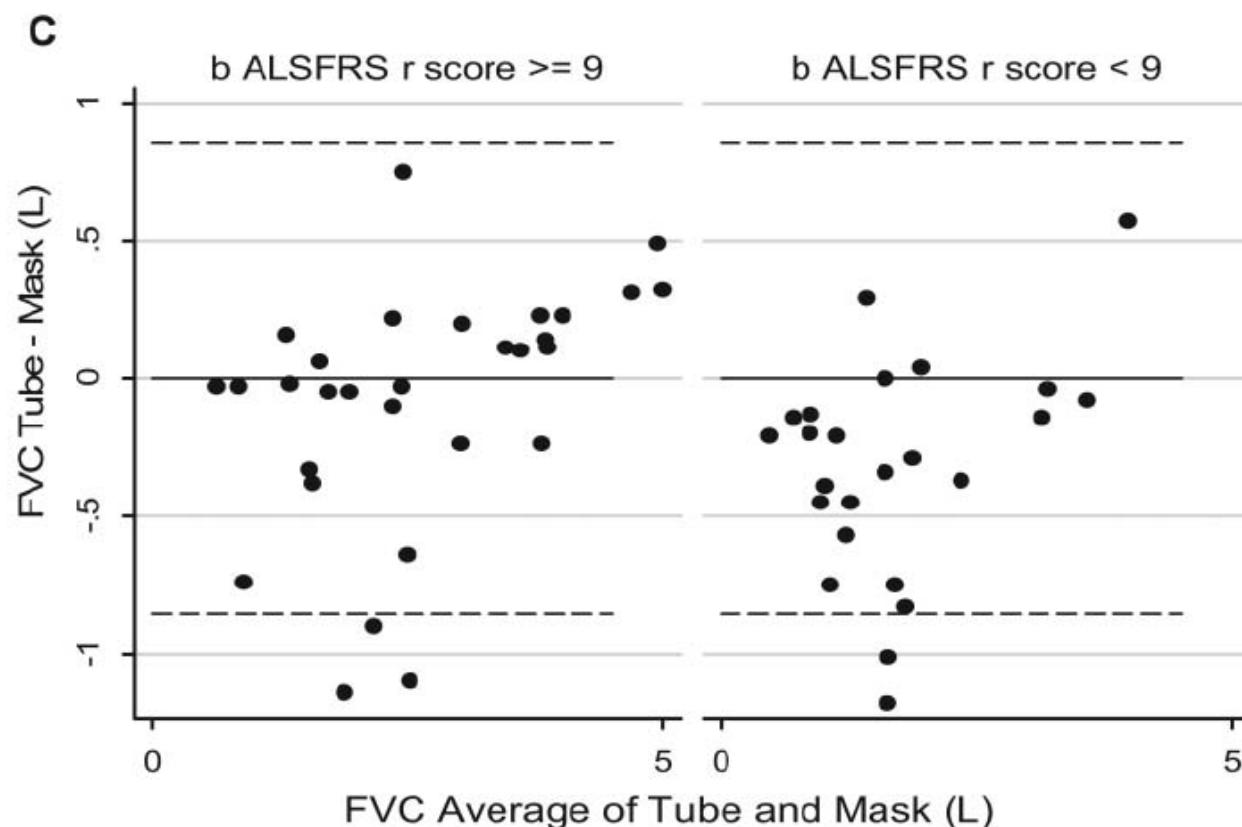
$$\text{PAO}_2 = 150 - (\text{PACO}_2 / r)$$

$$r = \text{VCO}_2 / \text{VO}_2 (> 0.8)$$

Observacionist: _____

The role of facemask spirometry in motor neuron disease

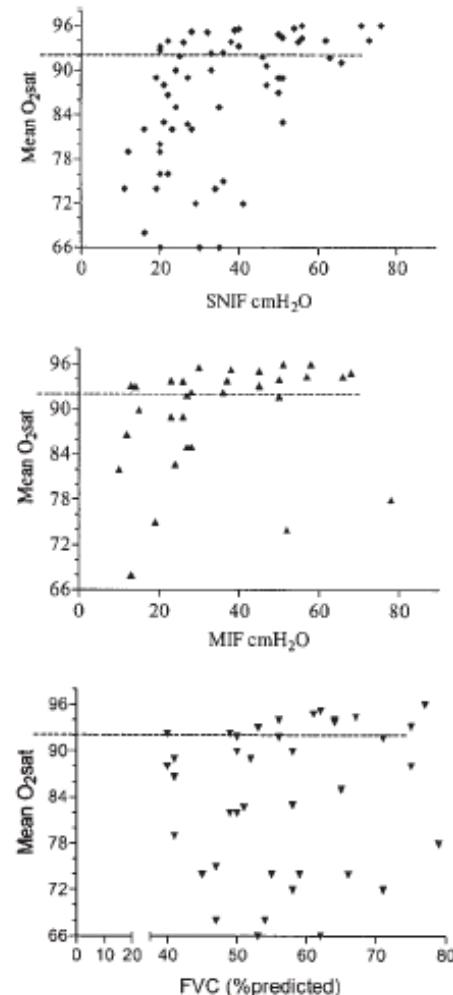
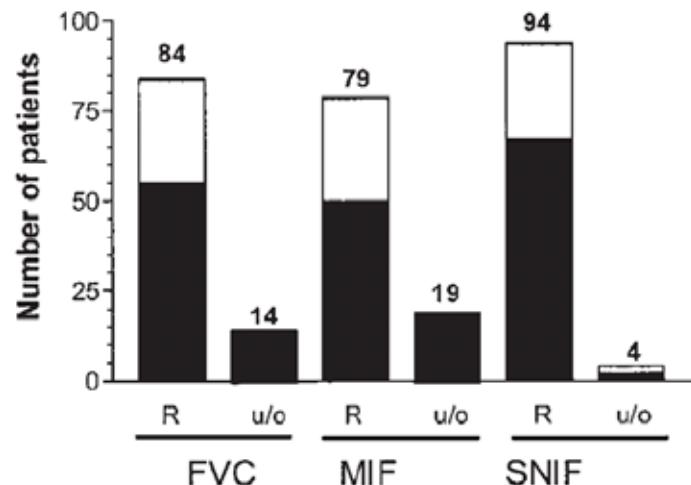
Thorax April 2013 Vol 68 No 4

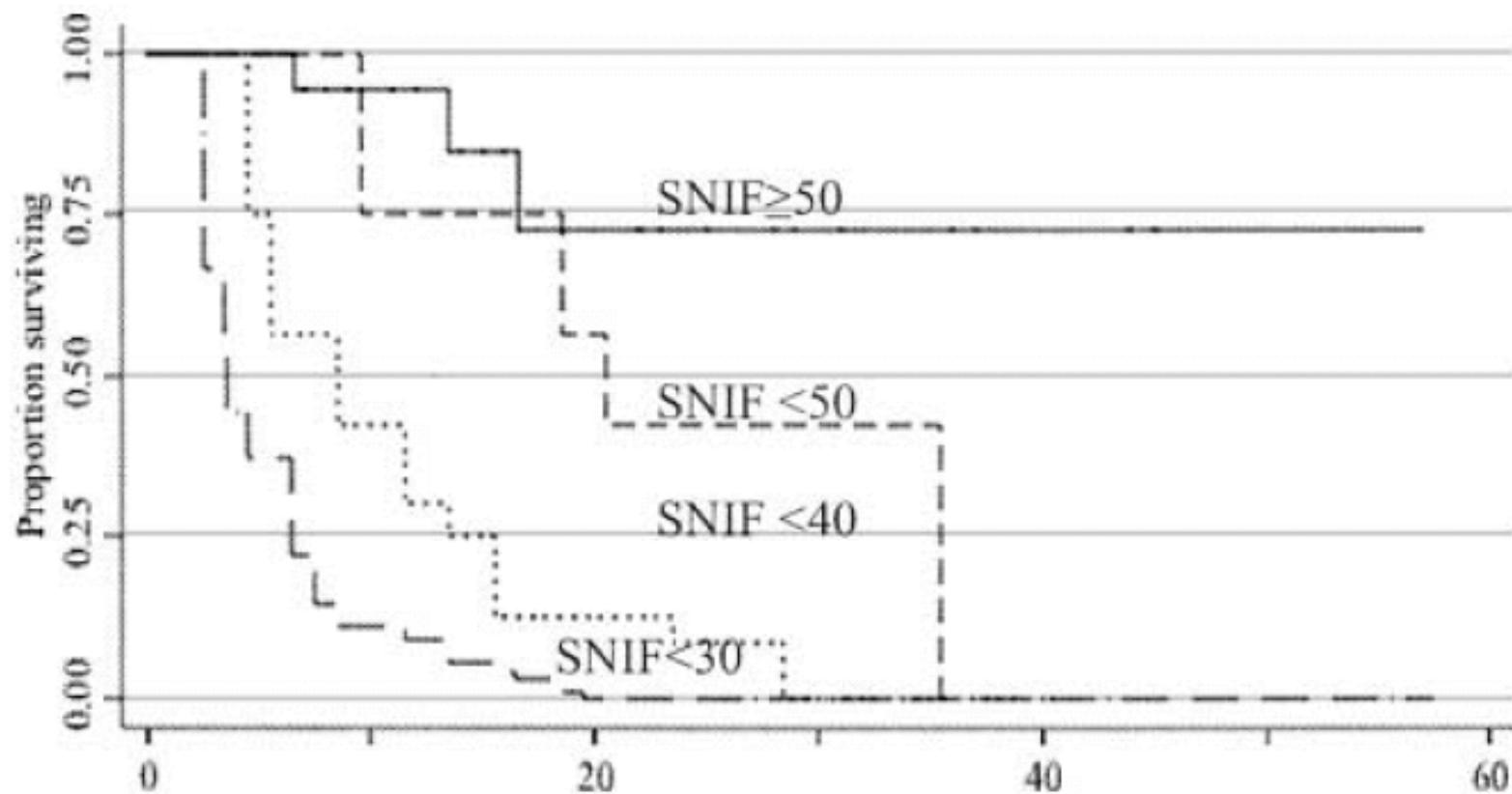


Use of Sniff Nasal-Inspiratory Force to Predict Survival in Amyotrophic Lateral Sclerosis

Ross K. Morgan*, Stephen McNally*, Michael Alexander, Ronan Conroy, Orla Hardiman, and Richard W. Costello

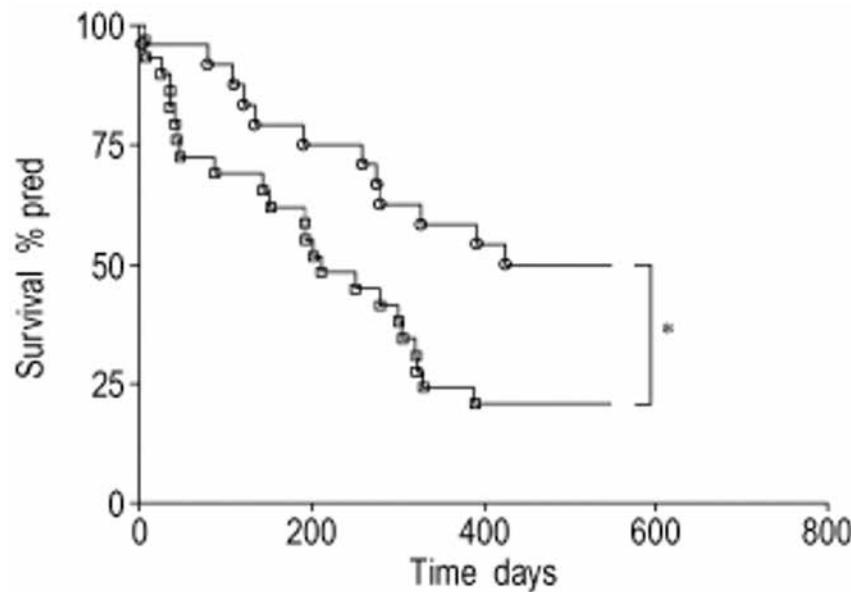
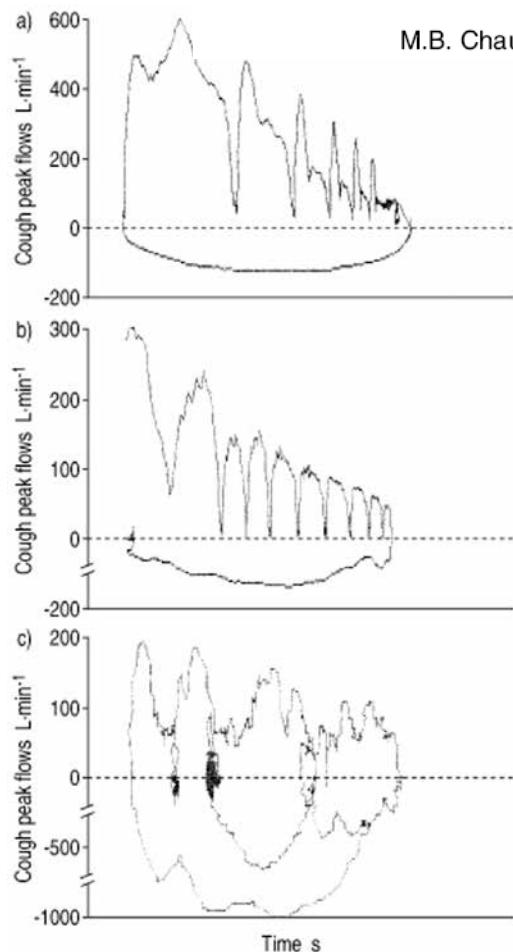
Departments of Medicine, Neurology, and Epidemiology, Royal College of Surgeons in Ireland,







Relationship between supramaximal flow during cough and mortality in motor neurone disease

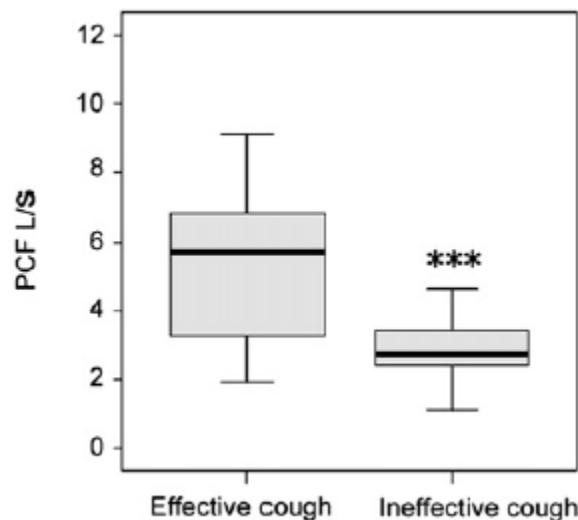


Predictors of Ineffective Cough during a Chest Infection in Patients with Stable Amyotrophic Lateral Sclerosis

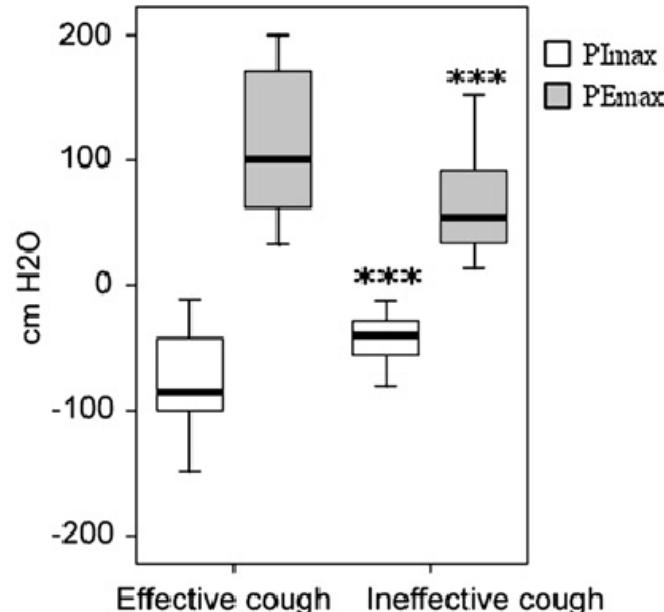
Jesús Sancho^{1,2}, Emilio Servera^{1,2}, Juan Díaz², and Julio Marín²

¹Respiratory Care Unit and ²Department of Respiratory Medicine, Hospital Clínico Universitario, Universitat de Valencia, Valencia, Spain

Am J Respir Crit Care Med Vol 175. pp 1266–1271, 2007



255 l/min



Inconvenients de la consulta multidisciplinar

- Temps de visita perllongat
- Pèrdua de intimitat
- Difusió de responsabilitats



EDITORIAL

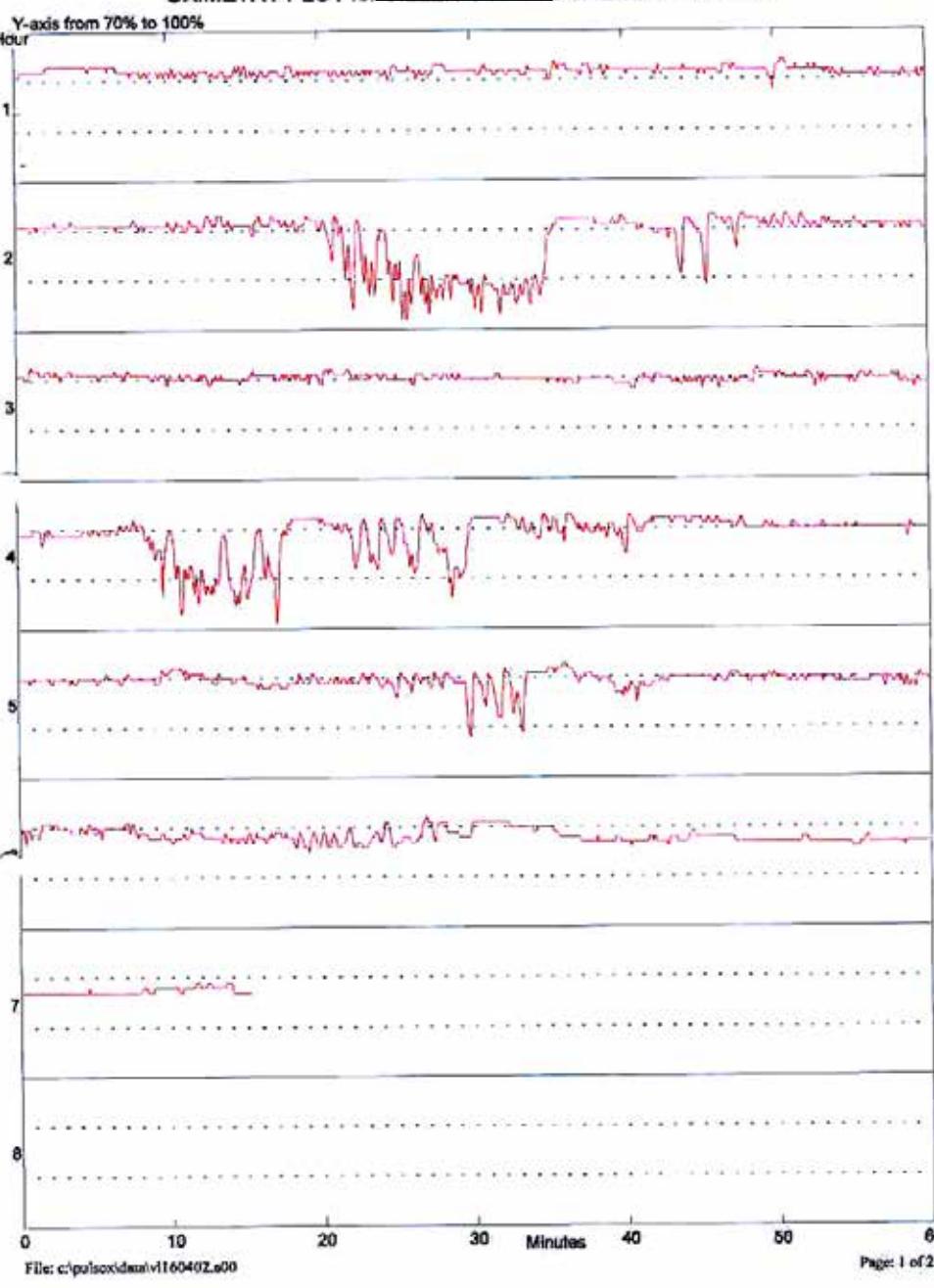
Daytime predictors of sleep-disordered breathing in neuromuscular patients to better schedule polysomnography

F. Lofaso*,[#], B. Fauroux^{†,+}, D. Orlikowski* and H. Prigent*

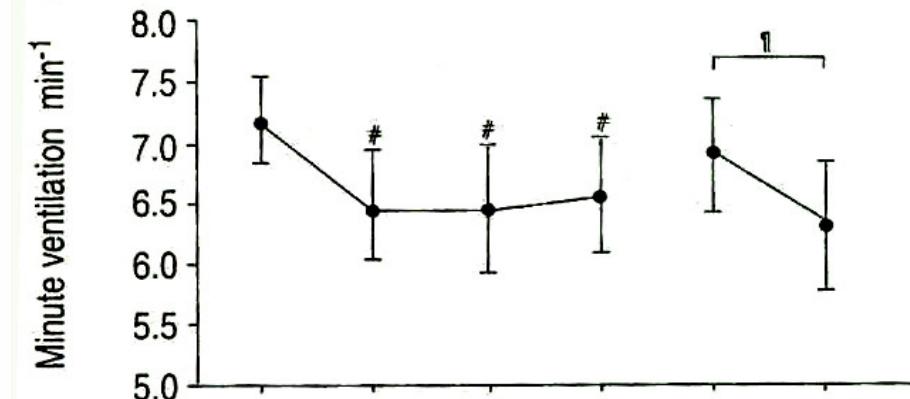
OXIMETRY PLOT for VILLENT CUBEL D

PULSOX SaO₂ Analysis Ver 2.0a

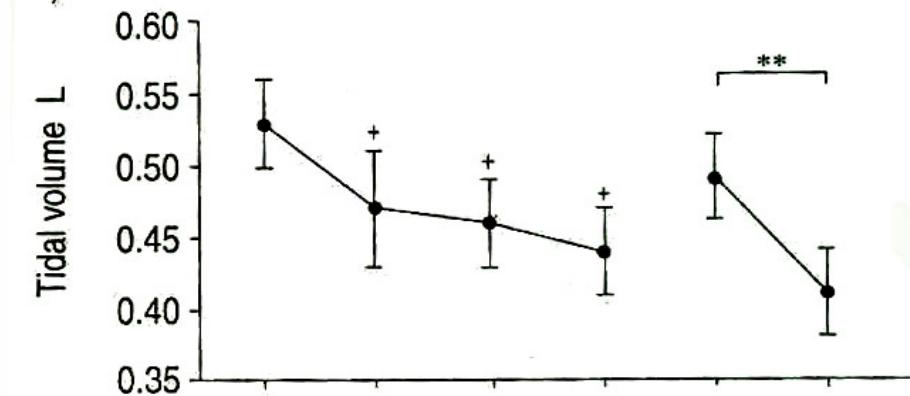
OXIMETRY PLOT for [REDACTED] D 3:12:00 16/04/2002



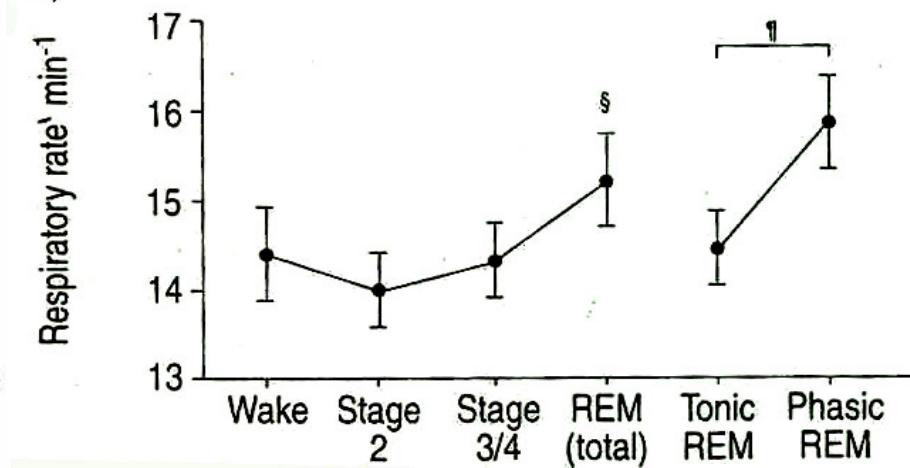
a)



b)



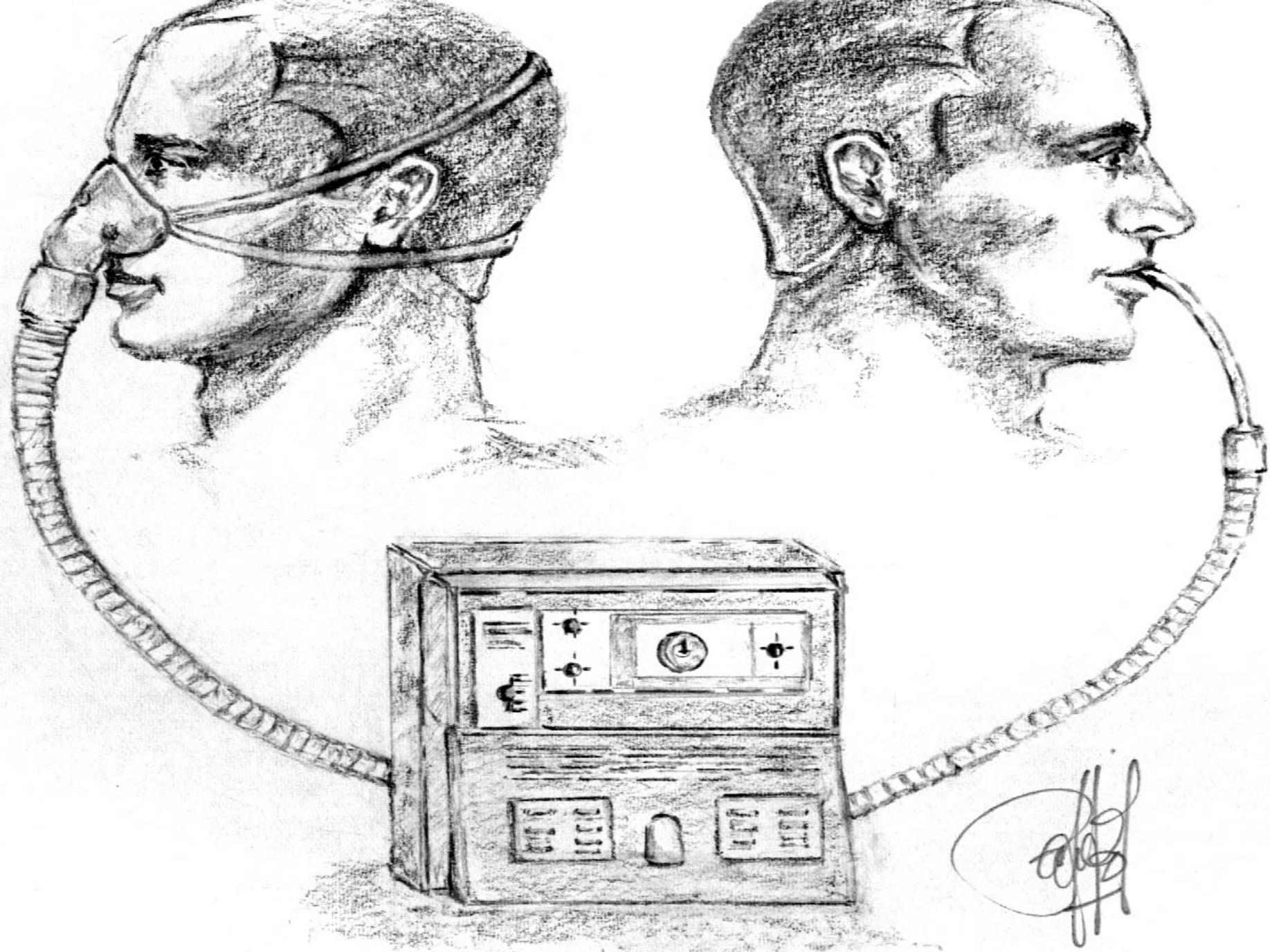
c)



Abordatje terapeutic precoç

Abordatge terapèutic precoç

- Decisió precoç (maneig invasiu i no invasiu)
- Fisioteràpia respiratòria precoç
- Moment ideal per iniciar la VNI
- Detecció precoç de problemes amb la VNI



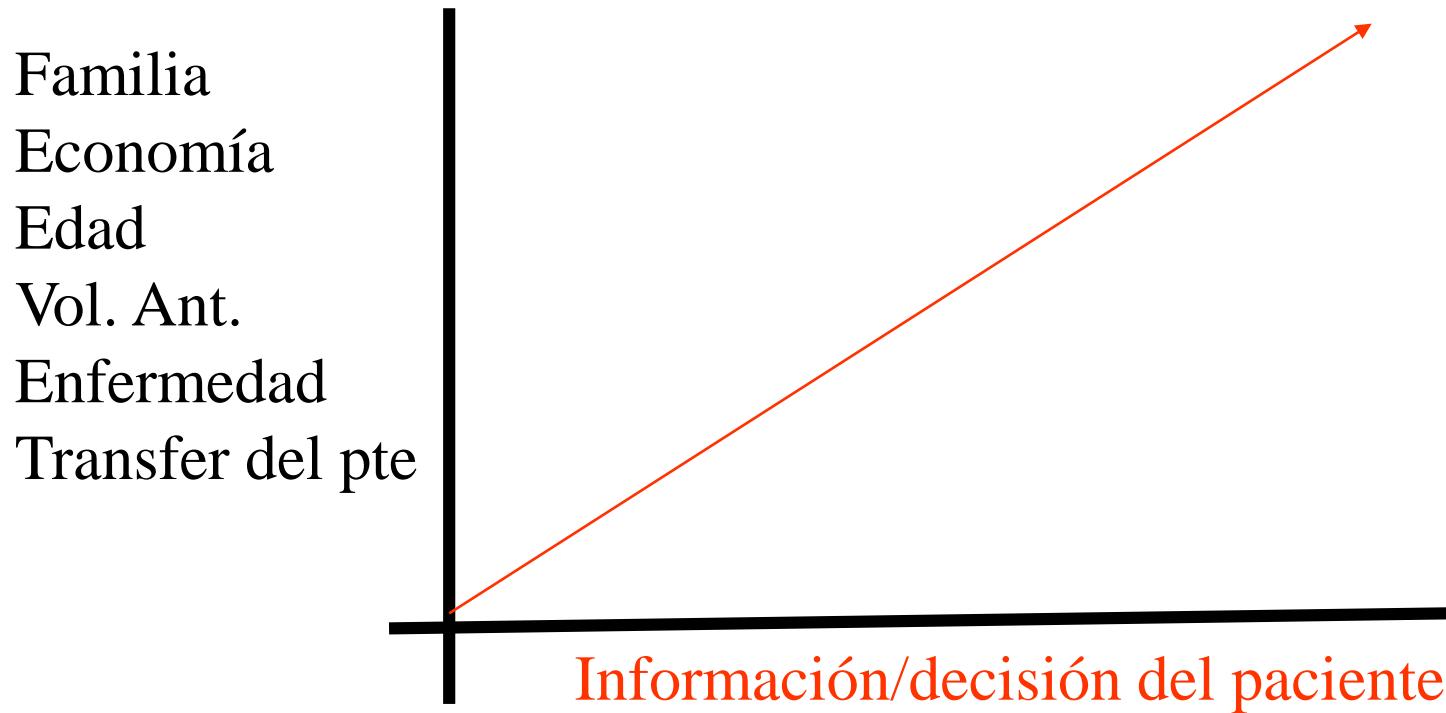
Stability of Choices about Life-sustaining Treatments

Marion Danis, MD; Joanne Garrett, PhD; Russell Harris, MD; and Donald L. Patrick, PhD

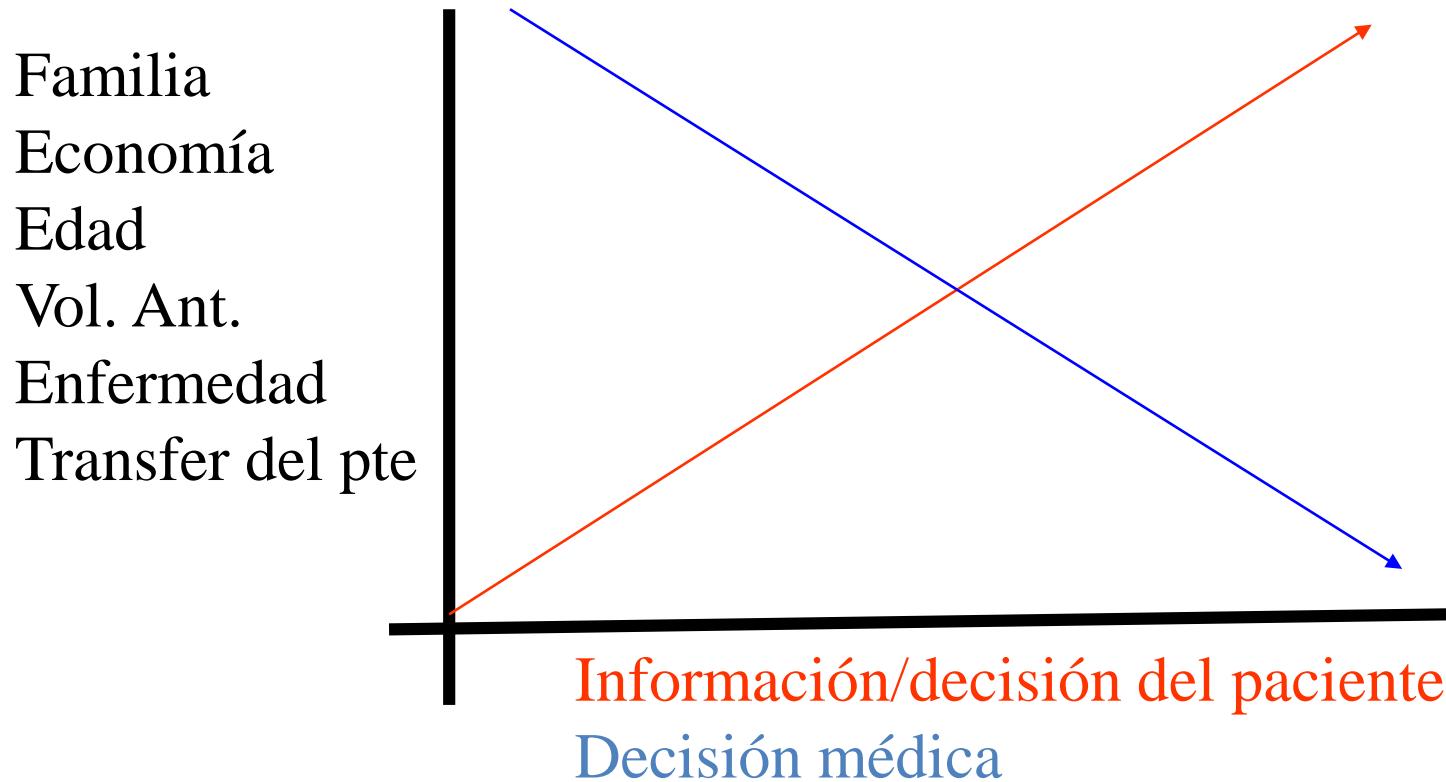
Ann Intern Med. 1994;120:567-573.

<i>¿desearía asistencia ventilatoria en un futuro? n=2536</i>	<i>Inicio</i>	<i>Evol, cambio de opinión</i>
Ventilación si	70 %	69%
Ventilación no	11%	12%

Grado de información y decisión



Grado de información y decisión





Outcome and attitudes toward home tracheostomy ventilation of consecutive patients: A 10-year experience

Santino Marchese^{a,*}, Daniele Lo Coco^b, Albino Lo Coco^a

^aPulmonary and Respiratory Intensive Care Unit, Ospedale Civico, ARNAS, Palermo, Italy

^bALS Research Center, Dipartimento di Neurologia, Oftalmologia e Psichiatria, University of Palermo, Italy

Received 16 June 2007; accepted 8 October 2007

Available online 19 November 2007

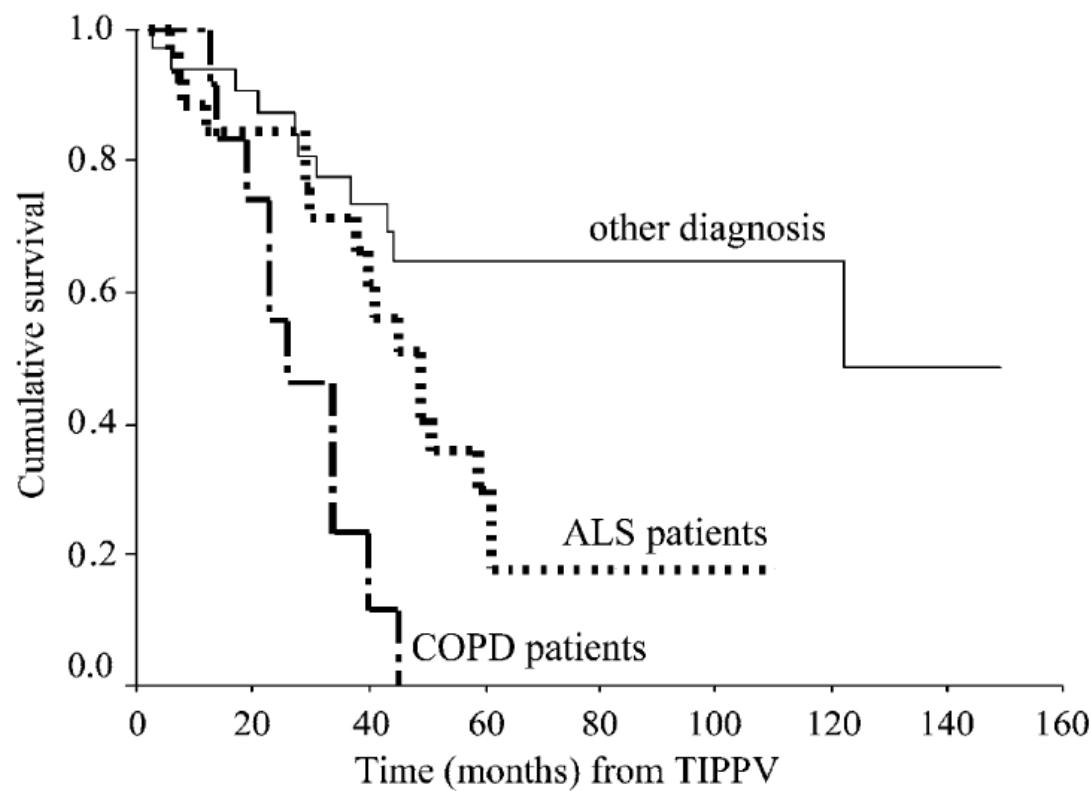
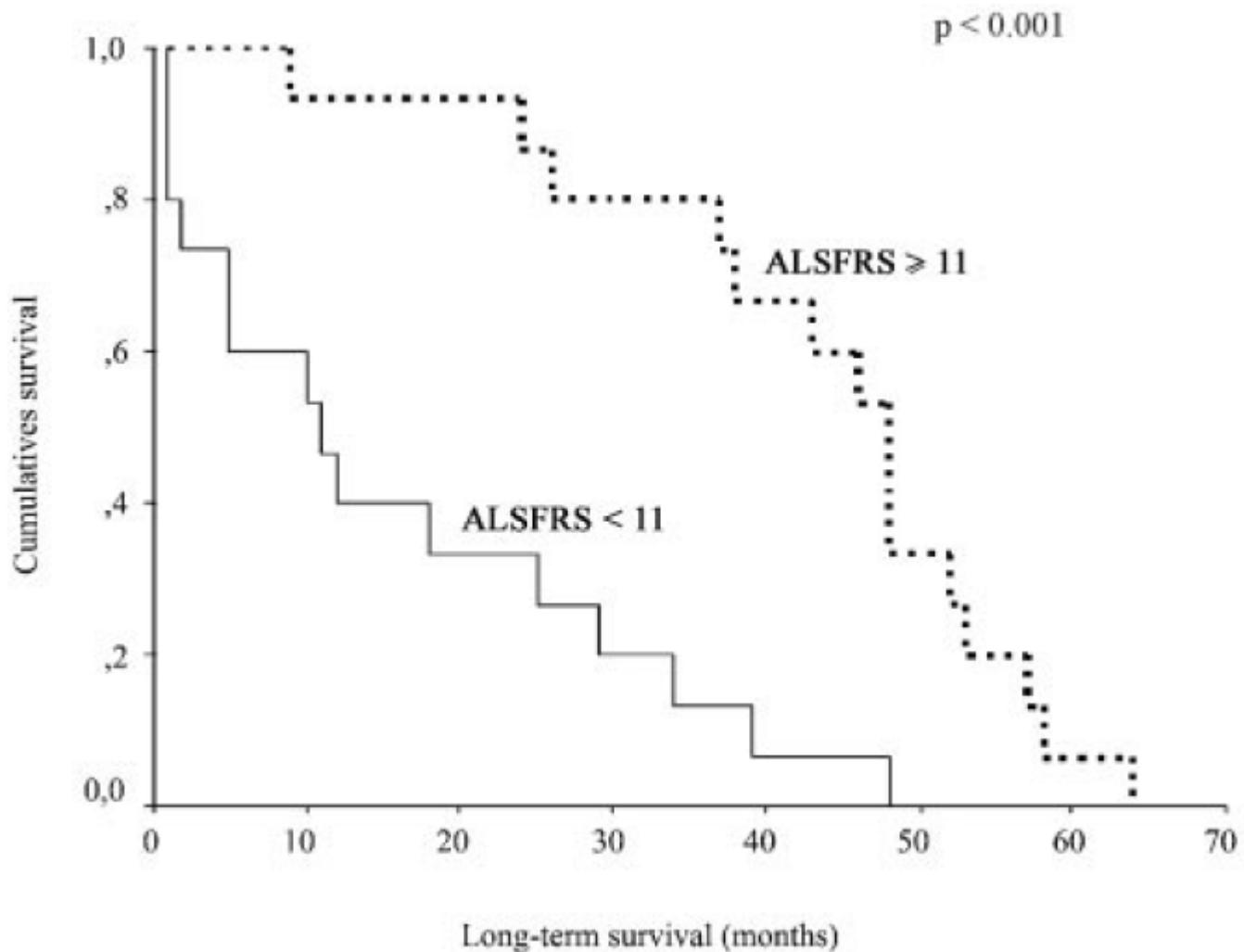


Figure 2 Comparison of survival between COPD patients ($n = 14$) ALS patients ($n = 30$) and the other diagnostic groups ($n = 33$). COPD and ALS patients showed a significant lower survival (COPD vs. other patients ($p = 0.0001$), ALS patients vs. other patients ($p = 0.02$)).



(CHEST 2007; 132:64–69)

PRESENCIA DE COMPLICACIONES RESPIRATORIAS

1. Criterios de ventilación mecánica: hipercapnia, desaturación arterial nocturna, síntomas (ortopnea) CVF < 50% del valor teórico.
2. Dificultad para el drenaje de secreciones: clínica compatible, PFT < 270 l/min.
3. Disfagia: clínica compatible, test diagnóstico.

Consensuar con el paciente y la familia

MANEJO NO INVASIVO

1. Ventilación no invasiva.
2. Asistencia a la tos (mecánica, manual).
3. Rehabilitación y medidas preventivas de aspiración.

MANEJO INVASIVO

1. Traqueostomía.
2. Gastrostomía percutánea.

Fracaso terapéutico,
considerar

SEGUIMIENTO EN CONSULTAS EXTERNAS, CONSULTA MULTIDISCIPLINAR
Y ATENCIÓN DOMICILIARIA ESPECIALIZADA
VALORAR TRATAMIENTO PALIATIVO

Fisioterapia precoç



CHEST

Original Research

PULMONARY REHABILITATION

Respiratory Muscle Training for Respiratory Deficits in Neurodegenerative Disorders

A Systematic Review

Alvaro Reyes, MSc; Mel Ziman, PhD; and Ken Nosaka, PhD

CHEST 2013; 143(5):1386–1394

Table 2—*Inspiratory Muscle-Training Studies*

Study/Year	Participants	Training Protocol	Training Load	Outcome Measures	Results
Sapienza et al ²⁵ /2011	60 patients with idiopathic Parkinson disease Disability level II to III Intervention: 30 25 male, 5 female Nonintervention: 30 22 male, 8 female	Intervention: 5 sets 5 repetitions 5 d/wk 4 wk Nonintervention Same protocol	Intervention: 75% average MEP Nonintervention: MEP never increased	FVC FEV ₁ PEF FEV ₁ /FVC	Intervention: 27% increase in MEP Nonintervention: 4% decrease in MEP
Troche et al ²⁶ /2010	60 patients with idiopathic Parkinson disease Disability level II to IV Intervention: 30 25 male, 5 female Nonintervention: 30 22 male, 8 female	Intervention: 5 sets 5 repetitions 5 d/wk 4 wk Nonintervention Same protocol	Intervention: 75% average MEP Nonintervention: No physiologic load	P/A score Swallowing timing Duration of hyoid elevation Quality of life	Intervention: Improved P/A score Improvement in laryngeal function during swallowing Improvement in swallowing-related quality of life Nonintervention: No significant changes
Pitts et al ²⁷ /2009	10 male patients with Parkinson disease Hoehn and Yahr II to III	5 sets 3 repetitions 5 d/wk 4 wk	75% average MEP	P/A score MEP PEF Cough volume acceleration	Significant increase in MEP Significant decrease in P/A score Increase in cough volume acceleration
Chiara et al ²⁸ /2006	17 patients with multiple sclerosis Mild to moderate disability: 17 3 male, 14 female Healthy participants: 14 2 male, 12 female	Intervention: 4 sets 6 repetitions 5 d/wk 8 wk Healthy participants: Same protocol	Intervention: 40% MEP 1st wk 60% MEP 2nd wk 80% MEP 3rd through 8th wk Healthy participants: Same protocol	FVC FEV ₁ PEF Maximal voluntary cough	Patients with multiple sclerosis: MEP increased PEF increased Improvement in cough airflow and cough volume occurred only in patients with moderate level of disability Healthy participants: MEP increased
Gosselink et al ²⁹ /2000	15 patients with multiple sclerosis Intervention: 9 3 male, 6 female Nonintervention: 9 6 male, 3 female	Intervention: 3 sets bid 15 repetitions 7 d/wk 3 mo Nonintervention: Breathing exercises	Intervention: 60% MEP Nonintervention: Not applicable	FVC MIP MEP Cough efficacy	Intervention: Cough efficacy significantly improved relative to control group MEP and MIP not statistically different from control group Nonintervention: No significant changes
Smeltzer et al ³⁰ /1996	15 patients with multiple sclerosis Eligible if baseline MEP's were between 45% to 60% of predicted MEP Intervention: 10 5 male, 5 female Nonintervention: 5 2 male, 3 female	Intervention: 3 sets bid 15 repetitions 7 d/wk 3 mo Nonintervention: Same protocol device configured to train inspiratory muscles	Intervention: Not reported Nonintervention: Not reported	MIP MEP	Intervention: Significant increased MEP Nonintervention: No significant changes

MEP = maximal expiratory pressure; MIP = maximal inspiratory pressure; P/A = penetration/aspiration; PEF = peak expiratory flow.

Table 3—Inspiratory Muscle-Training Studies

Study/Year	Participants	Training Protocol	Training Load	Outcome Measures	Results
Inzelberg et al ⁶ /2005	20 patients with Parkinson disease Disability level II and III Intervention: 10 male, 1 female Nonintervention: 10 male, 1 female	Intervention: 6 d/wk 30 min per session 12 wk Nonintervention: Same protocol	Intervention: 15% MIP first week 5%-10% increasing each session 60% MIP at end of the first month 60% at new MIP achieved Nonintervention: Low load	Spirometric indexes MIP Inspiratory muscle endurance Perception of dyspnea Quality of life	Intervention: Significant increase in MIP and inspiratory muscle endurance compared with control group Significant decrease in perception of dyspnea Nonintervention: No significant changes
Fry et al ⁸ /2007	41 patients with multiple sclerosis Intervention: 20 Nonintervention: 21	Intervention: 3 sets 15 repetitions 7 d/wk 10 wk Nonintervention: No treatment	Intervention: Increased from 30% MIP according to Borg RPE and symptoms Nonintervention: Not applicable	Spirometric indexes MIP MEP Maximal voluntary ventilation	Intervention: Improved FEV ₁ , FVC, and FEF _{25%-75%} Significant greater improvement than the control group in MIP Nonintervention: No significant changes
Klefbeck and Hamrah Nedjad ⁷ /2003	15 patients with multiple sclerosis EDSS score between 6.5 and 9.5 Intervention: 8 male, 1 female Nonintervention: 8 male, 5 female	Intervention: 3 sets 10 repetitions 7 d/wk 10 wk Nonintervention: Deep breathing exercises	Intervention: 40%-60% MIP Nonintervention: Not applicable	Spirometric indexes MIP MEP Borg RPE scale	Intervention: MIP and MEP increased Nonintervention: No significant changes
Cheah et al ³⁰ /2009	19 patients with amyotrophic lateral sclerosis Intervention: 9 male, 3 female Nonintervention: 10 male, 4 female	Intervention: 10 min training 3 times/d 7 d/wk 12 wk Nonintervention: Same protocol	Intervention: 15% of maximal SNIP 30% 2nd wk 45% 3rd wk 60% 4th through 12 wk Nonintervention: No load	FVC VC MIP SNIP	Intervention: No significant changes Nonintervention: No significant changes

EDSS = expanded disability status scale; FEF_{25%-75%} = forced midexpiratory flow; RPE = rating of perceived exertion; SNIP = sustained nasal inspiratory pressure; VC = vital capacity. See Table 2 legend for expansion of other abbreviations.

CONCLUSIONS

There is some evidence that RMT, by using pressure threshold devices, improves a number of respiratory function parameters in patients with Parkinson disease and multiple sclerosis. However, the number of studies and their quality are insufficient to conclude whether IMT or EMT is effective in improving pulmonary function in patients with neurodegenerative disorders of the CNS. From the studies analyzed in the current review, it is possible, however, to

The Effects of Intermittent Positive Pressure Breathing on Patients with Respiratory Muscle Weakness^{1,2}

A. DE TROYER and P. DEISSE

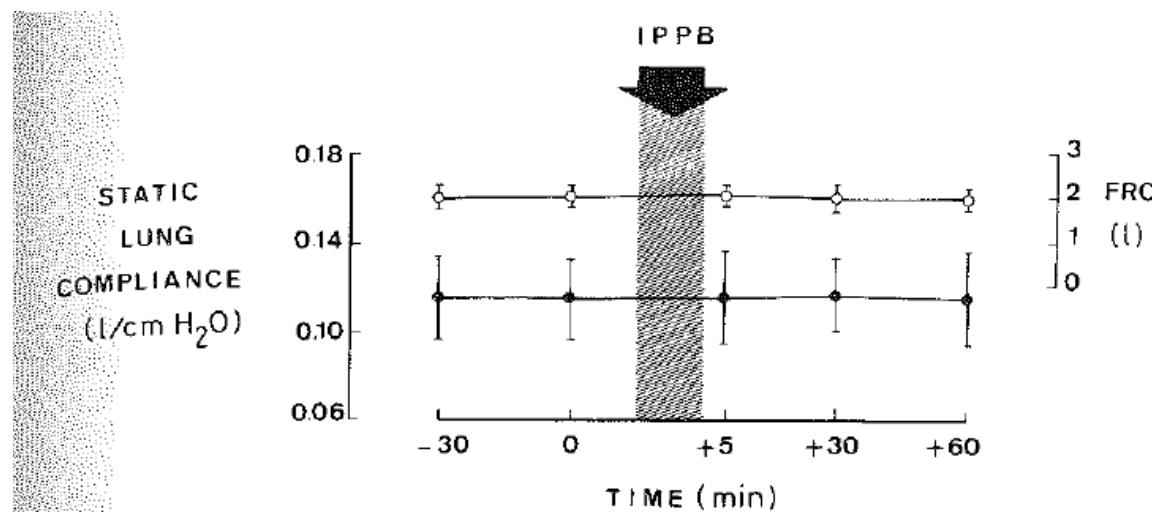


Fig. 3. The average changes in static expiratory lung compliance (closed circles) and functional residual capacity (FRC) (open circles) in 10 patients with respiratory muscle weakness after 15 min of intermittent positive pressure breathing (IPPB). The average Inflation pressure was 29 cm H₂O (range = 20 to 40 cm H₂O). Brackets indicate ± 1 SEM.

Cough augmentation in amyotrophic lateral sclerosis

N. Mustfa, MBBS; M. Aiello, MD; R.A. Lyall, MBBS; D. Nikoletou, MSc; D. Olivieri, MD; P.N. Leigh, PhD;
A.C. Davidson, MD; M.I. Polkey, PhD; and J. Moxham, MD

NEUROLOGY 2003;61:1285–1287

Table 2 Mean (SD) pressure and flow and flow changes achieved during maximal cough and assisted cough in nonbulbar patients

Cough assistance	TPP, cm H ₂ O	Pgas, cm H ₂ O	PCEF, L/min	% Increase in PCEF	Cough volume, L
Unassisted	49 (51)	75 (60)	217 (84)		1.16 (0.5)
Manually	65 (60)*	101 (71)†	244 (83)†	13	1.27 (0.6)
Exsufflation	66 (58)*	73 (63)	279 (87)†	28	
Insufflation	48 (55)	76 (63)	226 (86)	4	1.56 (0.5)†
Insufflation-exsufflation	63 (53)*	75 (60)	264 (73)†	21	

* $p < 0.01$, † $p < 0.001$.

TPP = transpulmonary pressure; Pgas = cough gastric pressure; PCEF = peak cough expiratory flow.



Aspiració no invasiva per TM ANITA



- 4. Tubuladura con filtro tipo Emstrong
- 5. Conector de 22x22mm
- 6. Conexión en L para ventilación por traqueo
- 7. Sondas de aspiración convencional
- 8. Fuente o sistema de aspiración

ANITA



Gentileza M. Segura

RESEARCH

Open Access

Effects of mechanical insufflation-exsufflation in preventing respiratory failure after extubation: a randomized controlled trial

Miguel R Gonçalves^{1,2*}, Teresa Honrado², João Carlos Winck¹ and José Artur Paiva²

Table 2 Postextubation outcomes data

	Group A (n = 40)	Group B (MI-E) (n = 35)
NIV application, n (%)	20 (50%)	14 (40%)
Reasons for NIV (n)		
Respiratory rate > 35 beats/min	5 (25%)	9 (64%)
SpO ₂ < 90%	4 (20%)	1 (7%)
20% variation of HR or BP	1 (5%)	-
PaO ₂ < 60; PaCO ₂ >45	10 (50%)	4 (29%)
Total period of MV (days)	17.8 ± 6.4 ^a	11.7 ± 3.5 ^a
Patients reintubated (n, %)	19 (48%) ^a	6 (17%) ^a
Causes of reintubation (n)		
Respiratory pauses with loss of consciousness	-	1
Respiratory distress after 2-h NIV	6	2
Decreasing level of consciousness	2	-
Intolerance to NIV	2	-
Hypotension (systolic BP < 90 mm Hg for > 30 minutes)	-	1
Secretion encumbrance associated with severe hypoxemia	9	2
NIV failure rate, n (%)	13 (65%) ^a	2 (14%) ^a
Total ICU length of stay	19.3 ± 8.1	16.9 ± 11.1
Postextubation ICU length of stay	9.8 ± 6.7 ^a	3.1 ± 2.5 ^a

Data are presented as mean ± standard deviation. APS II, New Simplified Acute Physiology Score; COPD, chronic obstructive pulmonary disease; MV, mechanical ventilation; NIV, noninvasive ventilation; NS, nonsignificant. ^aP < 0.05

VM precoç aventatges teòriques

- Millora de la mecànica respiratoria (pulmonar y toràcica)
- Reducció secundaria del treball respiratori
- Correcció de la hipoventilació inicial i la inestabilitat a la VAS

Randomised trial of preventive nasal ventilation in Duchenne muscular dystrophy

*Jean-Claude Raphael, Sylvie Chevret, Claude Chastang, Françoise Bouvet, for the French Multicentre Cooperative Group on Home Mechanical Ventilation Assistance in Duchenne de Boulogne Muscular Dystrophy**

Lancet 1994; **343:** 1600–04

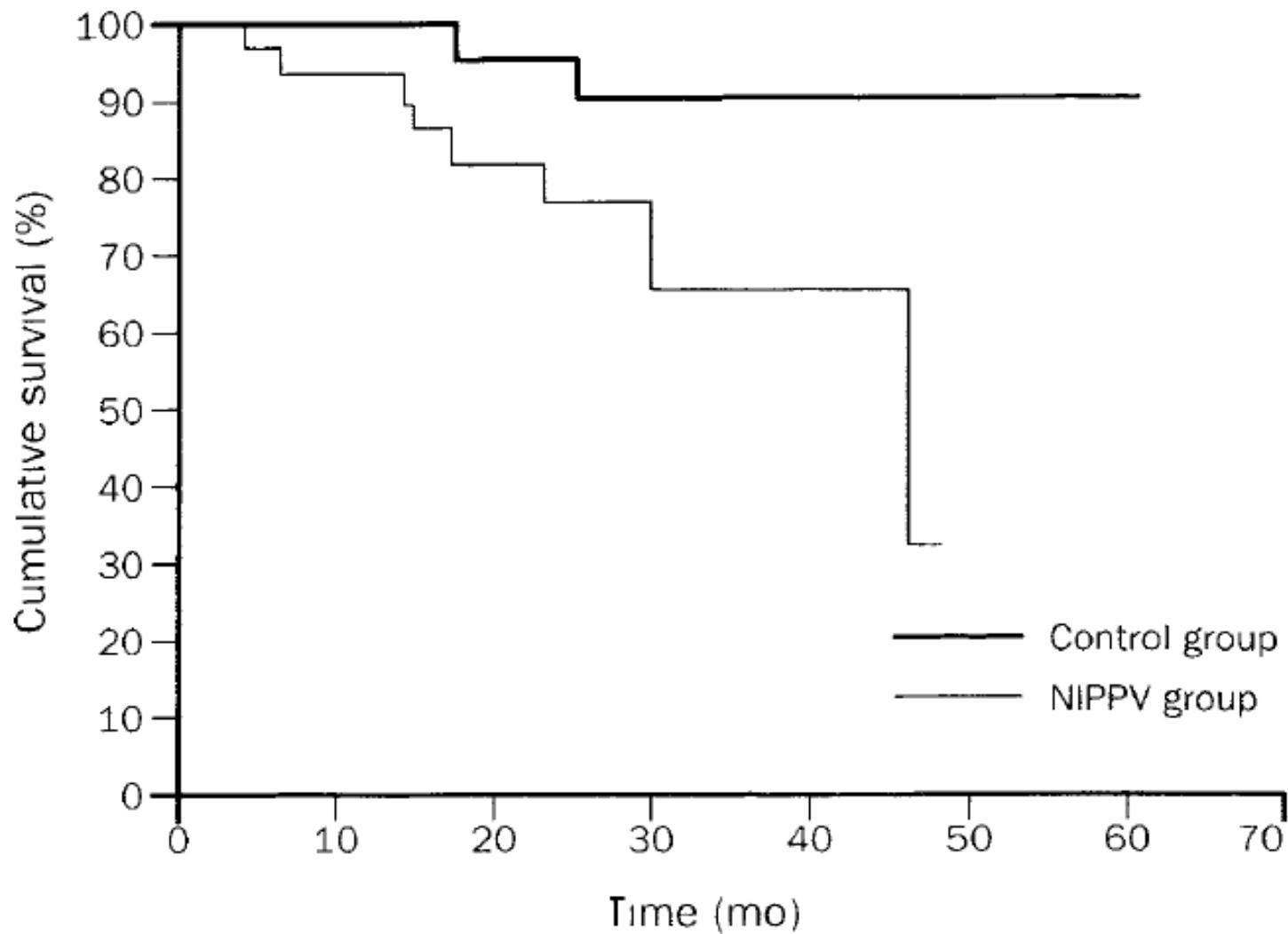


Figure 1: Kaplan-Meier plot of survival from randomisation in DMD patients receiving nocturnal NIPPV or conventional treatment

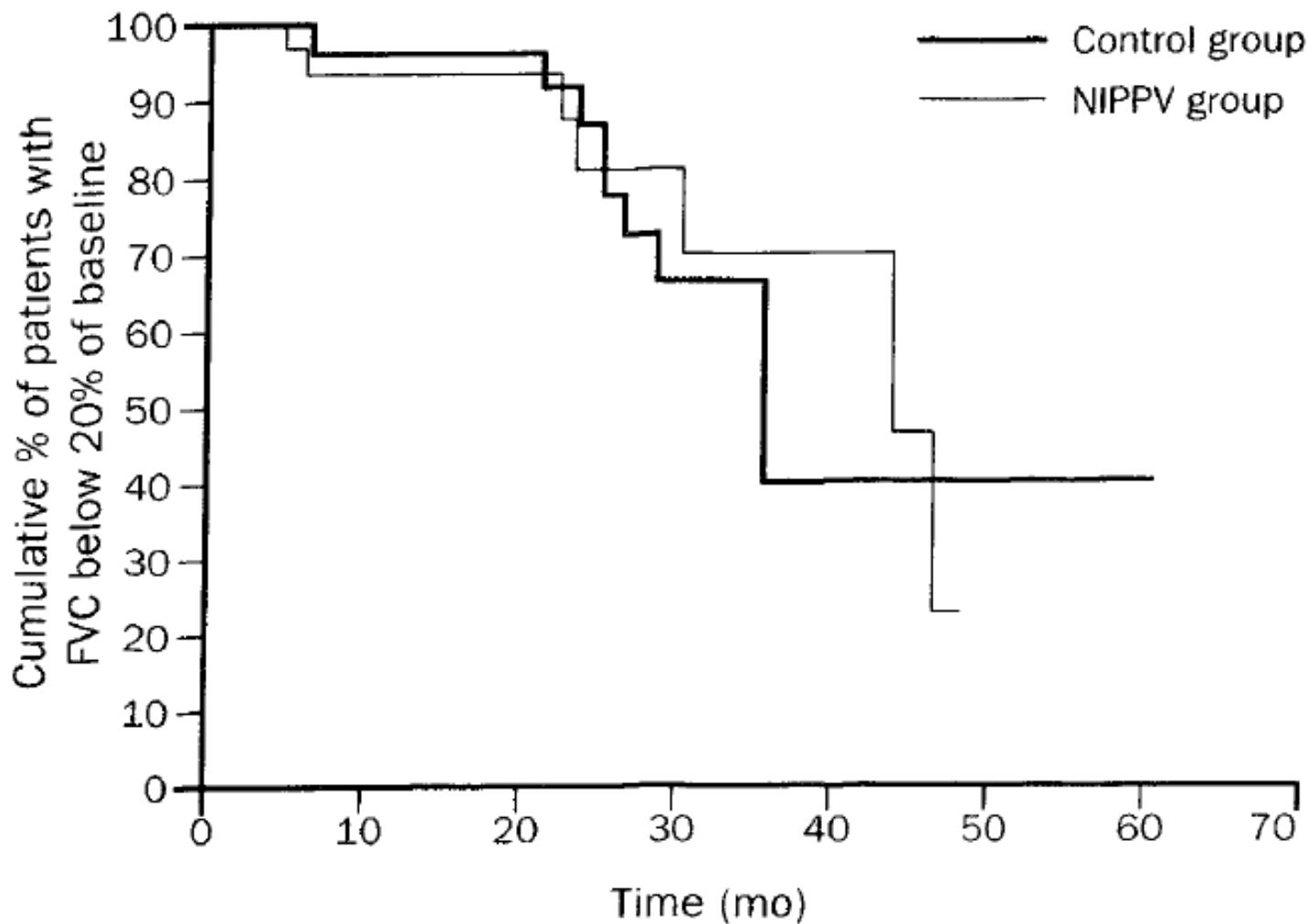


Figure 2: Estimated time to FVC below 20% of baseline in DMD patients receiving nocturnal NIPPV or conventional treatment

observed. Finally, a more realistic hypothesis to explain the higher mortality rate in the NIPPV group would be the false security that is associated with its use. Monitoring may have become less careful in the NIPPV group. This possibility is suggested by the lower frequency of necessary ventilated patients in the NIPPV group (9% vs 26% in the control group), whereas the rate of patients who fulfilled criteria for necessary ventilation was similar in the two groups (NIPPV 37% and control 34%). Such a bias could have been avoided by a double-blind controlled trial, but this is not a realistic possibility.

MECHANICAL VENTILATION

Randomised controlled trial of non-invasive ventilation (NIV) for nocturnal hypoventilation in neuromuscular and chest wall disease patients with daytime normocapnia

S Ward, M Chatwin, S Heather, A K Simonds

Thorax 2005;60:1019–1024. doi: 10.1136/thx.2004.037424

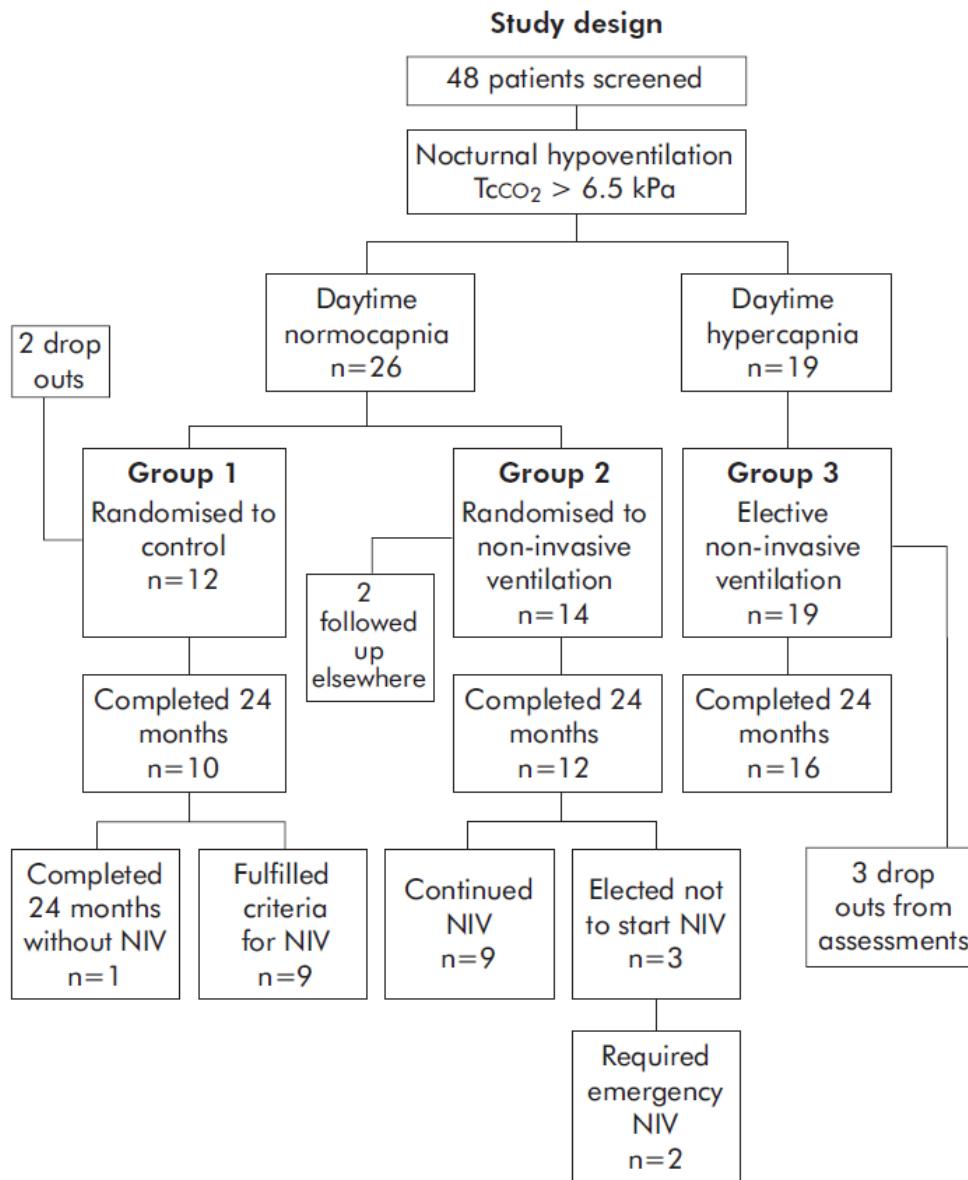
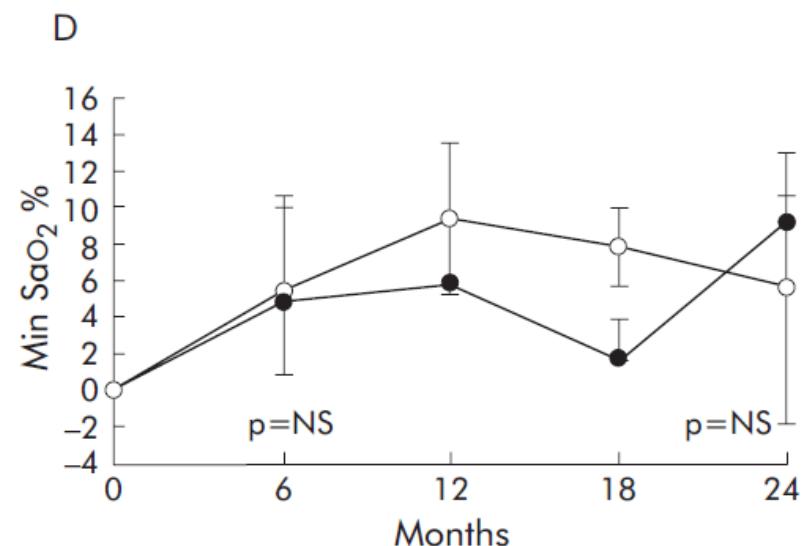
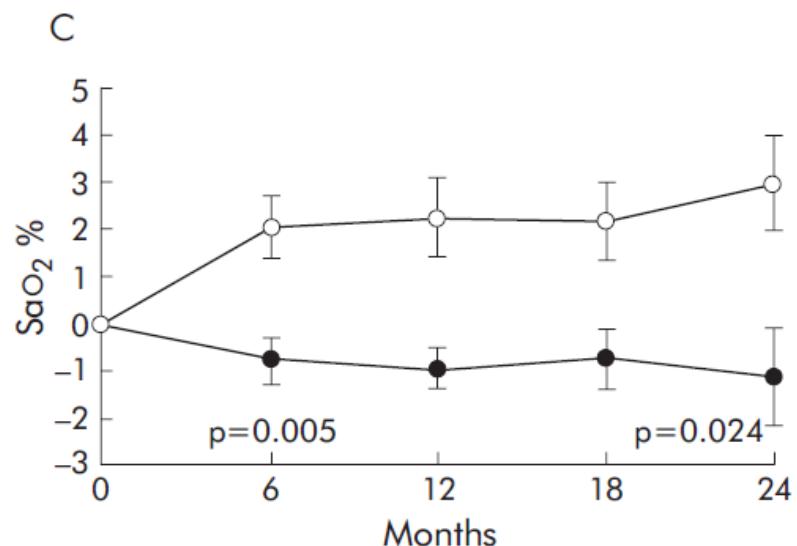
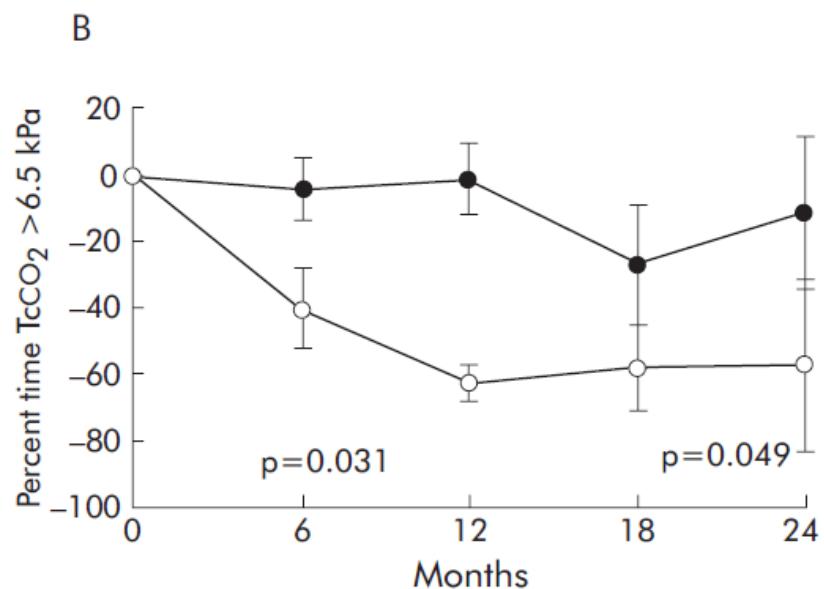
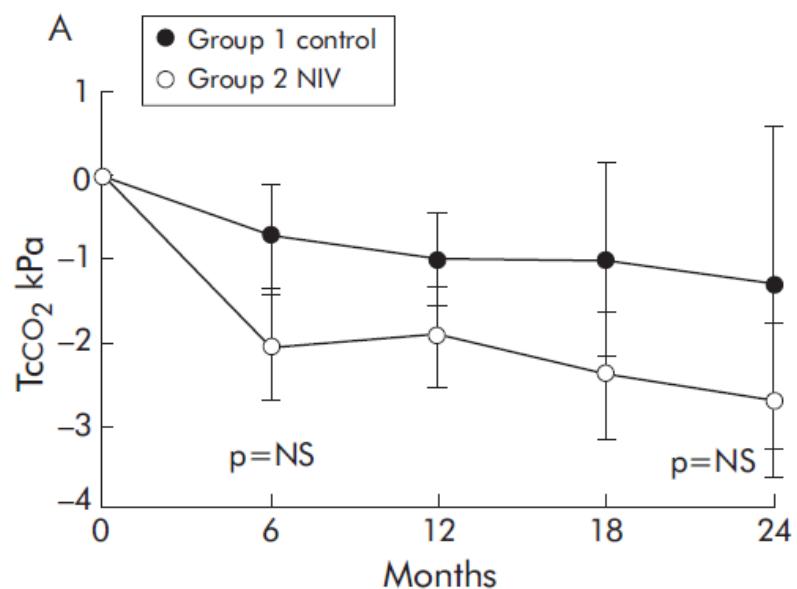


Figure 1 Study design.



Ward, et al

Thorax 2005; 60: 1019-1020

- La majoria del malalts del grup control (1) desenvolupen hipoventilació diürna en 24m
- La VNI NO millora ni l'estatus funcional respiratori ni la qualitat de vida dels malalts
- No hi ha diferència de mortalitat als 24m



25th International Symposium on ALS/MND

5 -7 December 2014
Brussels, Belgium

ABSTRACT SUBMISSION

Title: EFFECTS OF EARLY NON INVASIVE VENTILATION ON PULMONARY FUNCTION IN ALS PATIENTS: PRELIMINARY RESULTS OF A RANDOMIZED CONTROL TRIAL

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Abstract No. 0283

Title EFFECTS OF EARLY NON INVASIVE VENTILATION ON PULMONARY FUNCTION IN ALS PATIENTS:
PRELIMINARY RESULTS OF A RANDOMIZED CONTROL TRIAL

Abstract **BACKGROUND:** Non invasive ventilation has demonstrated an improvement of survival and quality of life in ALS patients. Recent studies (1,2) suggest that early initiation, in early stages of respiratory muscle weakness, would involve a greater increase in survival

OBJECTIVE: To assess the effects from early use of non invasive ventilation (NIV) in progression of respiratory muscle weakness, measured by rate of decline in Forced Vital Capacity (FVC).

METHODS: A multicentric, randomized, open-label, controlled clinical trial including patients with FVC < 75% who are randomized to: 1; early NIV (treatment initiation after randomization) and 2; standard NIV (treatment initiation when FVC < 50% predicted, presence of orthopnoea, and/or PaCO₂ > 45 mmHg). Patients were follow-up every three months. Variables collected included: anthropometric variables and data of ALS disease, functional respiratory variables (spirometry, arterial blood gases and nocturnal pulseoximetry).

Statistical Analysis

Non parametric test were used to compare groups at baseline. Differences in the FVC decline between groups over consecutive visits were compared using U Mann-Whitney and Wilcoxon test was used to analyze differences within each of the two groups.

RESULTS: 41 patients have been included; for the purpose of this study we analysed 25 patients (10 patients in group early NIV and 15 patients in standard group), 52 % men, mean age 59 (12) years and in the 92 % of cases the onset of the disease was limb. There weren't differences between both groups in anthropometric variables, data of ALS disease, arterial blood gases or nocturnal pulseoxymetry values. During the follow-up we only observed a slight significant decrease in FVC at 6 months in the early NIV group (FVC baseline 69%, FVC 3 months 69%, FVC 6 months 62% p=0.01) while there was a highly significant and progressive decrease in the standard group (FVC baseline 68%, FVC 3 months 53% p=0.000, FVC 6 months 45% p=0.009). Differences between groups were significant at 3 and 6 months.

CONCLUSIONS

Preliminary results in our study shows highly significant benefit effect of early NIV in ALS patients, slowing-down the progressive decline of Functional Vital Capacity.

→ Effects of non-invasive ventilation on survival and quality of life in patients with amyotrophic lateral sclerosis: a randomised controlled trial

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

Summary

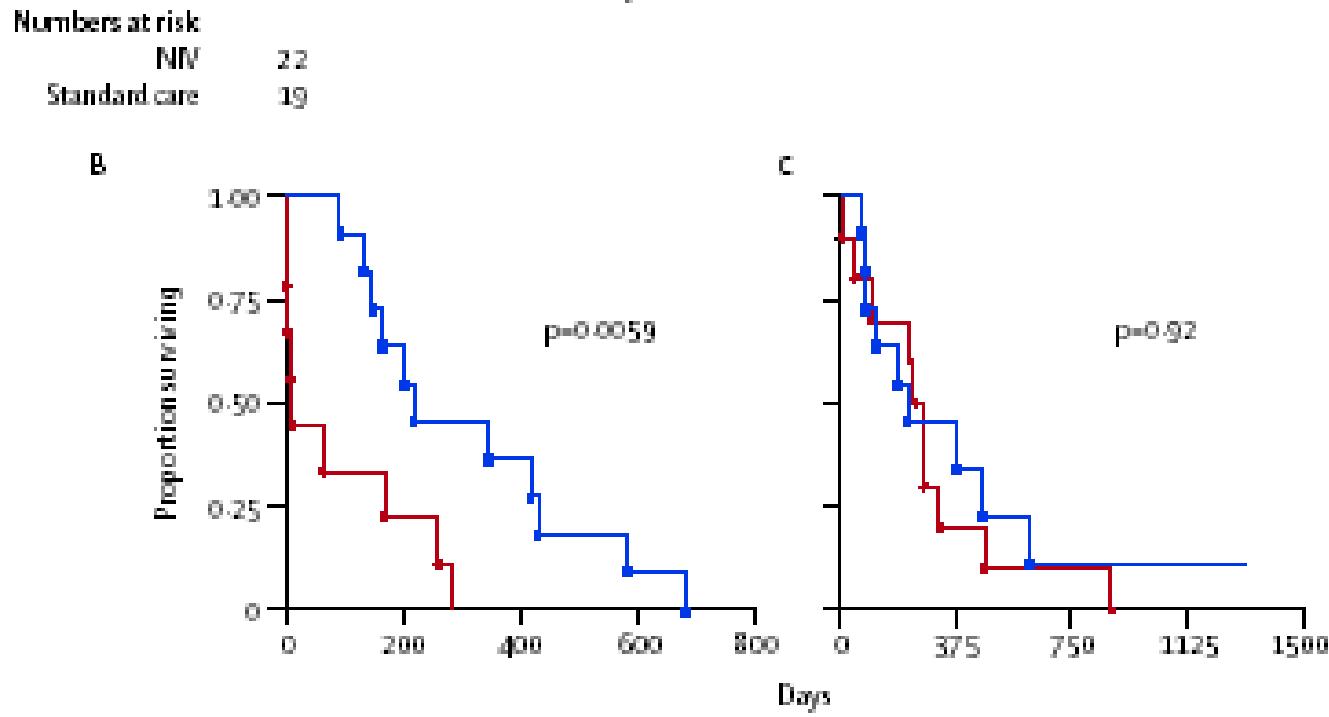
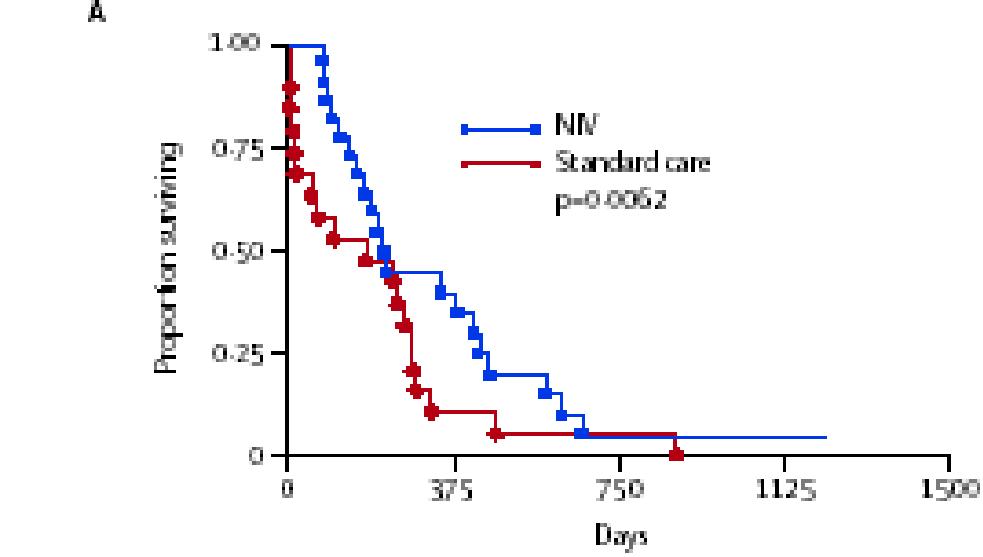
Background Few patients with amyotrophic lateral sclerosis currently receive non-invasive ventilation (NIV), reflecting clinical uncertainty about the role of this intervention. We aimed to assess the effect of NIV on quality of life and survival in amyotrophic lateral sclerosis in a randomised controlled trial.

Patients were subsequently randomly assigned to NIV or standard care only if and when they met one or both of the predefined criteria: orthopnoea with $P_{\text{CO}_2\text{max}}$ less than 60% of that predicted or symptomatic daytime hypercapnia.¹³ Random allocation was computer

Lancet Neurol 2006; 5: 140–47

Published online January 9, 2006

DOI:10.1016/S1474-4422(05)70326-4



Tractament Precoç?

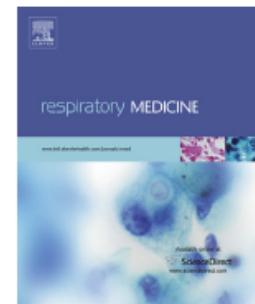
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Ambulatory adaptation to noninvasive ventilation in restrictive pulmonary disease: A randomized trial with cost assessment

Mercedes Pallero ^{a,b,c,1}, Carme Puy ^d, Rosa Güell ^d,
Caridad Pontes ^e, Sergi Martí ^{a,b,c,*}, Ferran Torres ^e,
Antonio Antón ^d, Xavier Muñoz ^{a,b,c,f}



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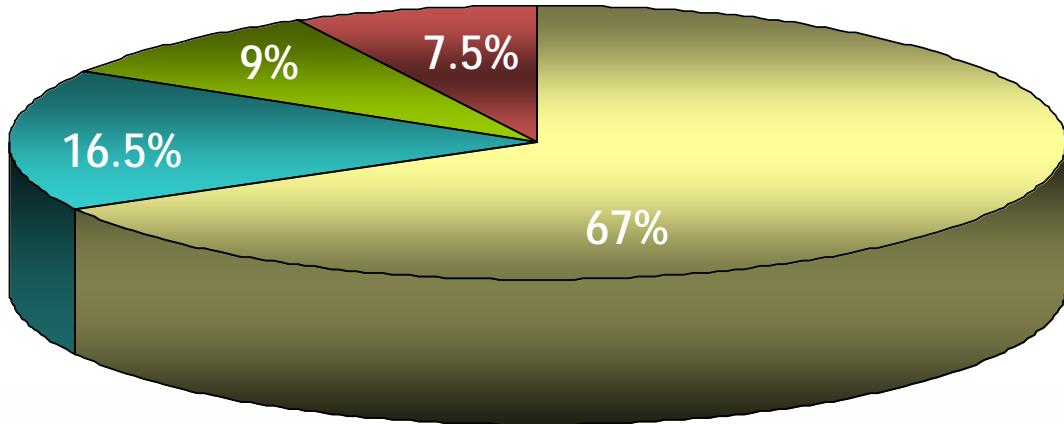


ORGANIZACIÓN DE LAS VISITAS





DESCRIPCION DE LOS PACIENTES POR SU TIPOLOGIA (n=188)



- **Patología Restrictiva**
- **Patología obstructiva**
- **Enfermedad Neromuscular de Larga Evolución**
- **ELA**

Conclusions

- El diagnòstic precoç de les complicacions respiratòries es fonamental
- El tractament multidisciplinar es desitjable
- La fisioteràpia respiratòria s'ha de iniciar lo mes abans possible per prevenir complicacions futures
- El tractament precoç amb VNI por ser una eina terapèutica amb futur

