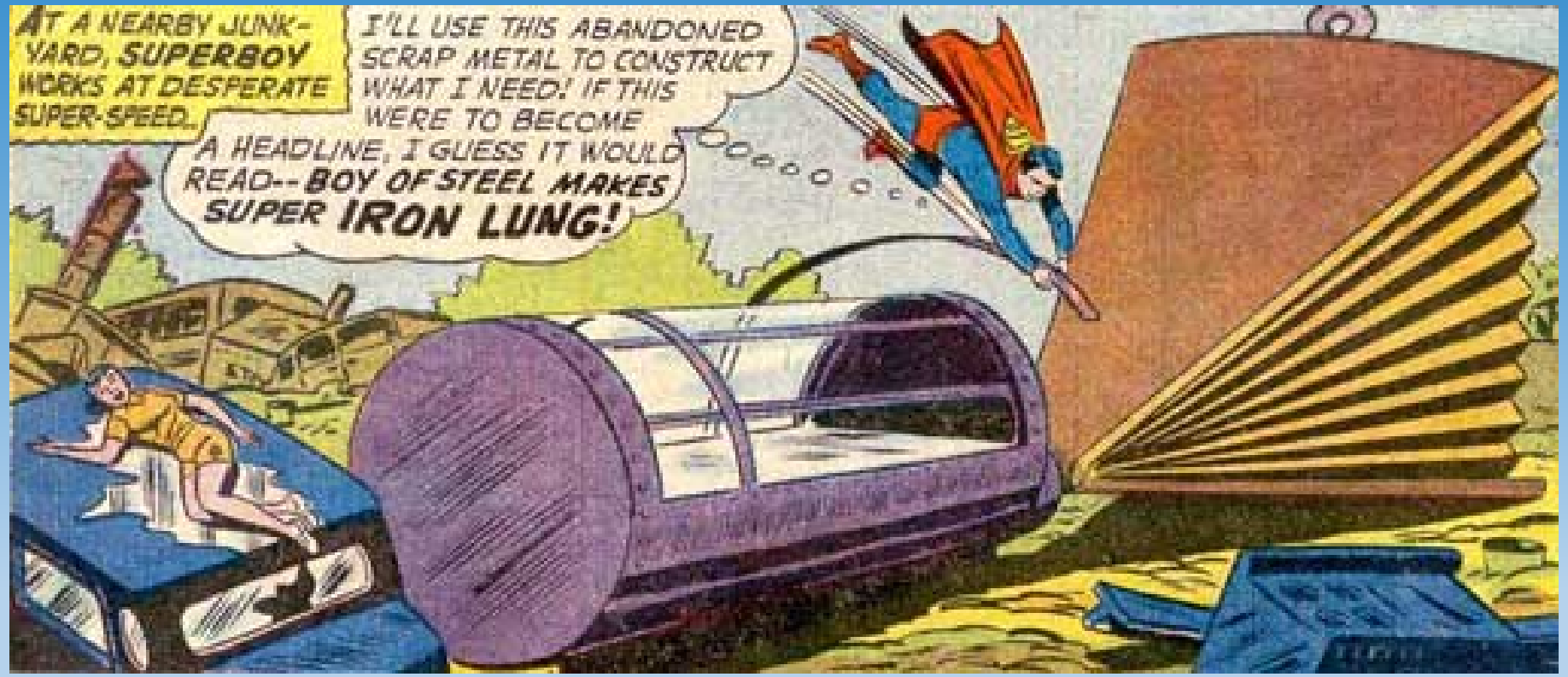
Prise en charge en kinésithérapie respiratoire des patients atteints de pathologies neuromusculaires: moyens et techniques à disposition des libéraux face aux limites en présence

Pierre LE CAM

Service de Réanimation Médicale et d'Assistance Respiratoire.

Hôpital de la Croix-rousse, GHN, Lyon

AKCR 2017


AT A NEARBY JUNK-YARD, SUPERBOY WORKS AT DESPERATE SUPER-SPEED..
I'LL USE THIS ABANDONED SCRAP METAL TO CONSTRUCT WHAT I NEED! IF THIS WERE TO BECOME A HEADLINE, I GUESS IT WOULD READ-- BOY OF STEEL MAKES SUPER IRON LUNG!

Les objectifs de la prise en charge respiratoire

La prise en charge respiratoire s'appuie sur les bilans respiratoires. Elle s'adapte aux besoins de la personne. Elle est réajustée lorsque la situation évolue. La prise en charge respiratoire s'intègre dans la prise en charge pluridisciplinaire (orthopédique, cardiologique, chirurgicale…).

OBJECTIFS

- Limiter les conséquences de l'atteinte musculaire sur la fonction respiratoire
- Limiter les facteurs aggravants
- Compenser le déficit des muscles respiratoires

Pour se sentir mieux au quotidien

Préserver la pompe respiratoire

- Maintenir ses propriétés mécaniques (mobilité, souplesse…)
- Favoriser la croissance des poumons et du thorax

→ **Kinésithérapie respiratoire**

→ **Hyperinsufflations, ventilation mécanique**

Désencombrer l'appareil respiratoire pour dégager les voies respiratoires

→ **Désencombrement bronchique : drainage bronchique et toux assistée**

Aider à respirer (restaurer la ventilation)

→ **Ventilation mécanique assistée :**
- **ventilation non invasive (VNI)**
- **ventilation invasive (par trachéotomie)**

Quality of Life, Physical Disability, and Respiratory Impairment in Duchenne Muscular Dystrophy

AJRCCM 2005

Malcolm Kohler, Christian F. Clarenbach, Lukas Böni, Thomas Brack, Erich W. Russi, and Konrad E. Bloch

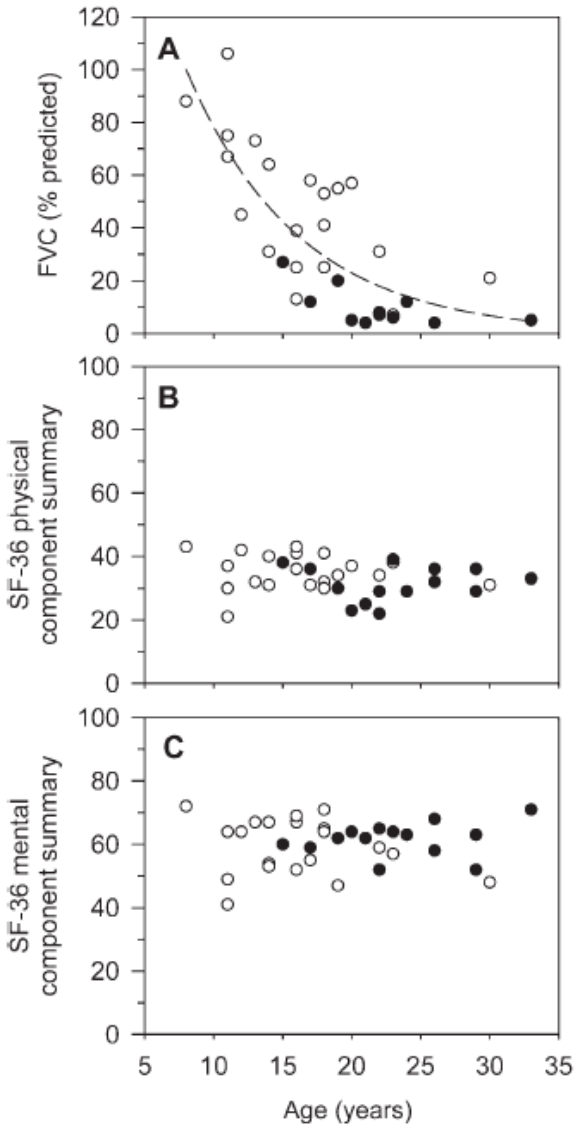


Figure 3. Respiratory impairment, represented by FVC values, progresses with advancing age. (*A*) The *dashed line* represents an exponential decay function ($f = a \cdot e^{-b \cdot time}$). There is little variation in the physical as well as in the mental component summaries of the SF-36 questionnaire (*B* and *C*, respectively). *Open circles*, patients without NIPPV; *closed circles*, patients with NIPPV.

35 pt (8-33 ans)

VNI = 14

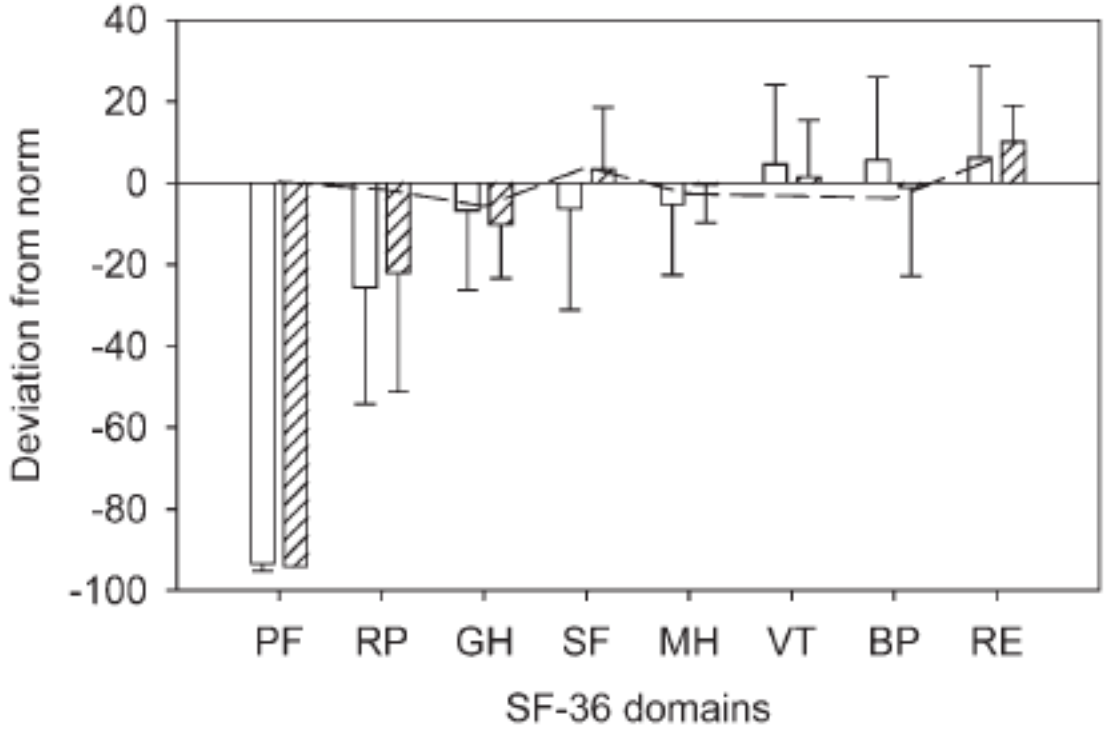


Figure 2. For each of the Short-Form 36 (SF-36) domains, the mean deviation (and SD) from sex- and age-matched U.S. reference values is displayed for 21 patients with Duchenne muscular dystrophy (DMD) without NIPPV (*open bars*), and for 14 patients with NIPPV (*hatched bars*). The *dashed line* represents the deviations of a German male reference population (20) from the U.S. reference (18). BP = bodily pain; GH= general health; MH = mental health; PF = physical functioning; RE = role-emotional; RP = role-physical; SF = social functioning; VT = vitality.

Kohler *AJRCCM* 2005

physiopathologie

Sleep and breathing in neuromuscular disease

Eur Respir J 2002; 19: 1194–1201

S.C. Bourke, G.J. Gibson

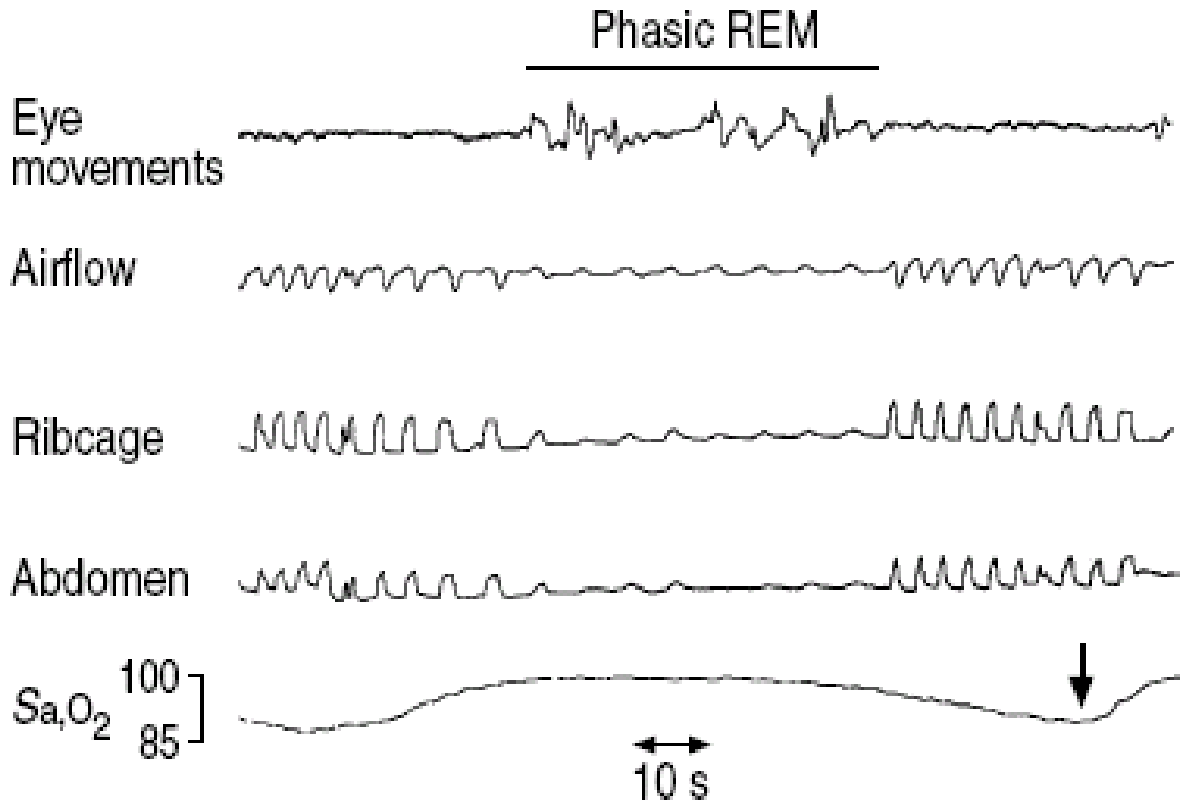
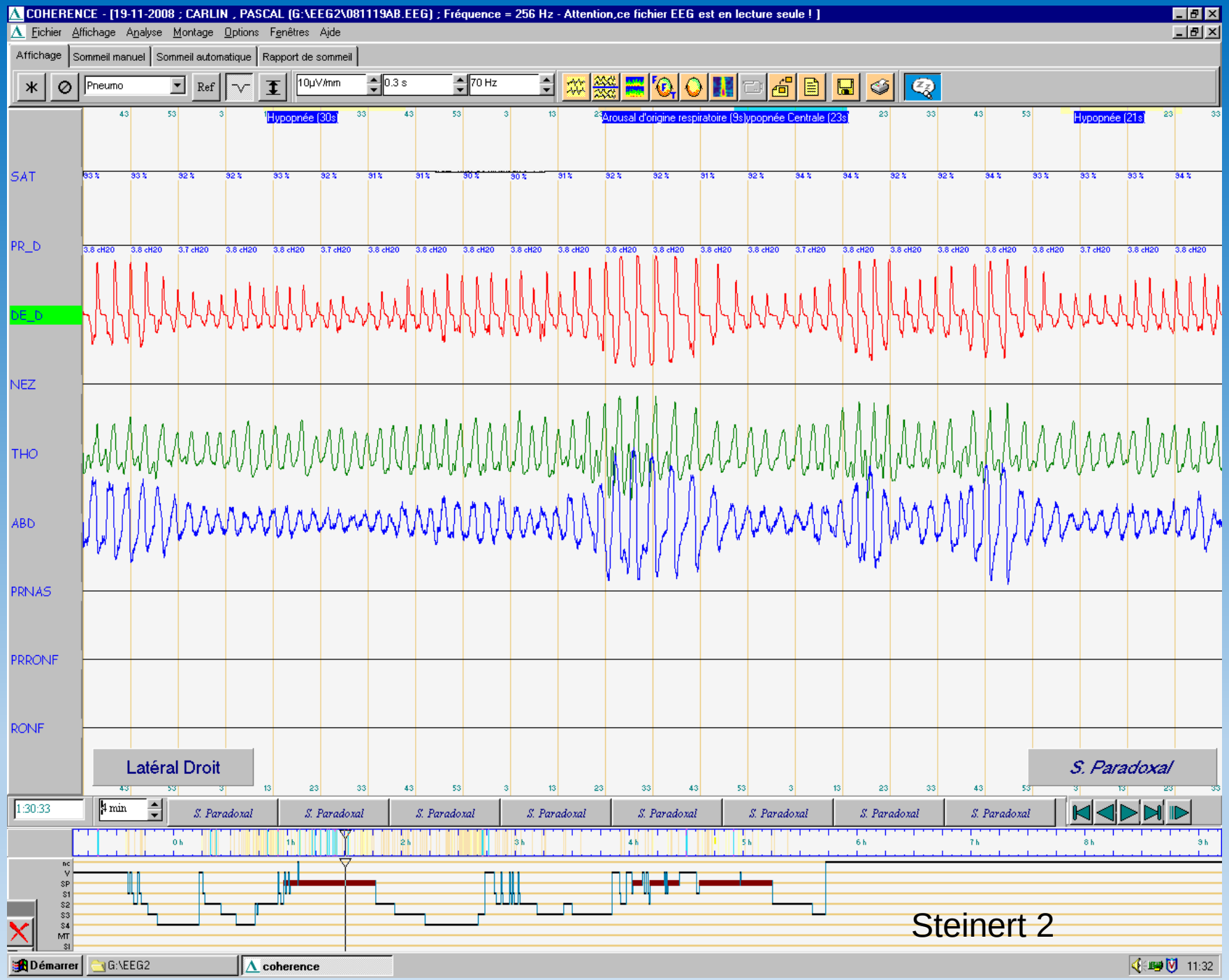


Fig. 2.–Suppression of airflow, chest and abdominal movement during tonic and phasic rapid eye movement (REM). Phasic REM is identified by the burst of rapid eye movements, and indicated by a bar. The consequent reduction in the arterial oxygen saturation (Sa,O_2) is indicated by the arrow.


COHERENCE - [19-11-2008 ; CARLIN , PASCAL (G:\EEG2\081119AB.EEG) ; Fréquence = 256 Hz - Attention,ce fichier EEG est en lecture seule !]
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Affichage Sommeil manuel Sommeil automatique Rapport de sommeil
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Ref
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0.3 s
70 Hz
Hypopnée [30s]
Arousal d'origine respiratoire [9s]
Hypopnée [21s]
SAT
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coherence
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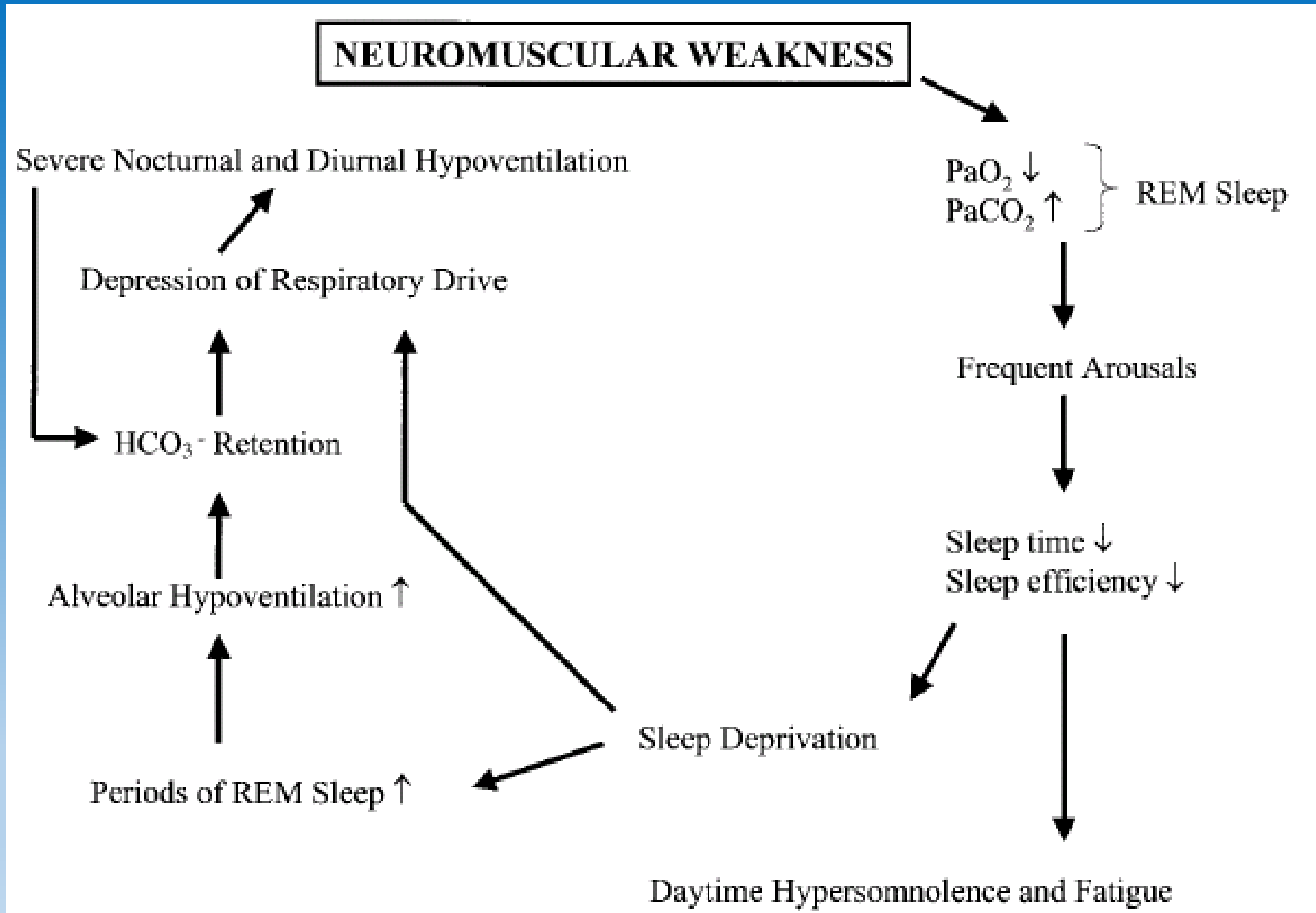


FIGURE 3. Circular events leading to severe hypoventilation in neuromuscular disease.

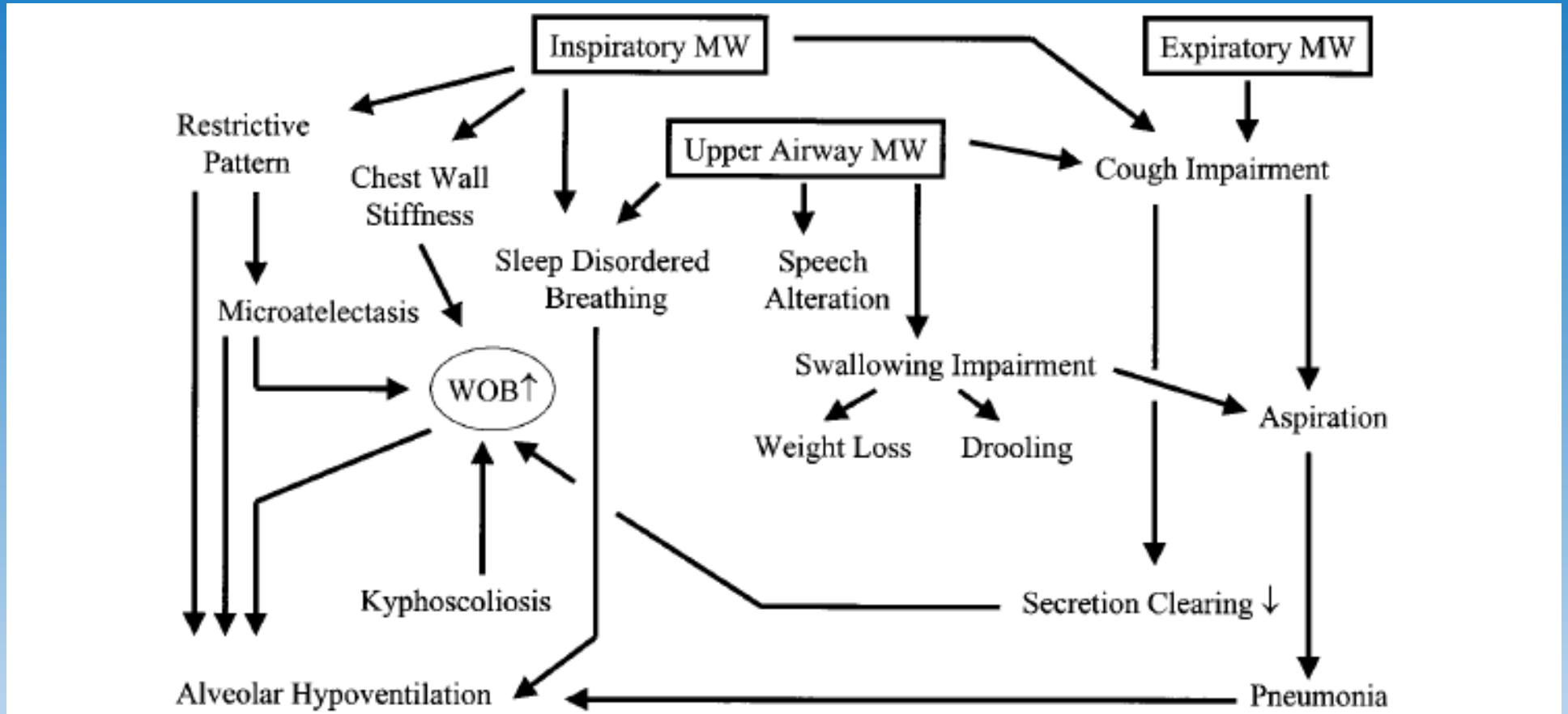
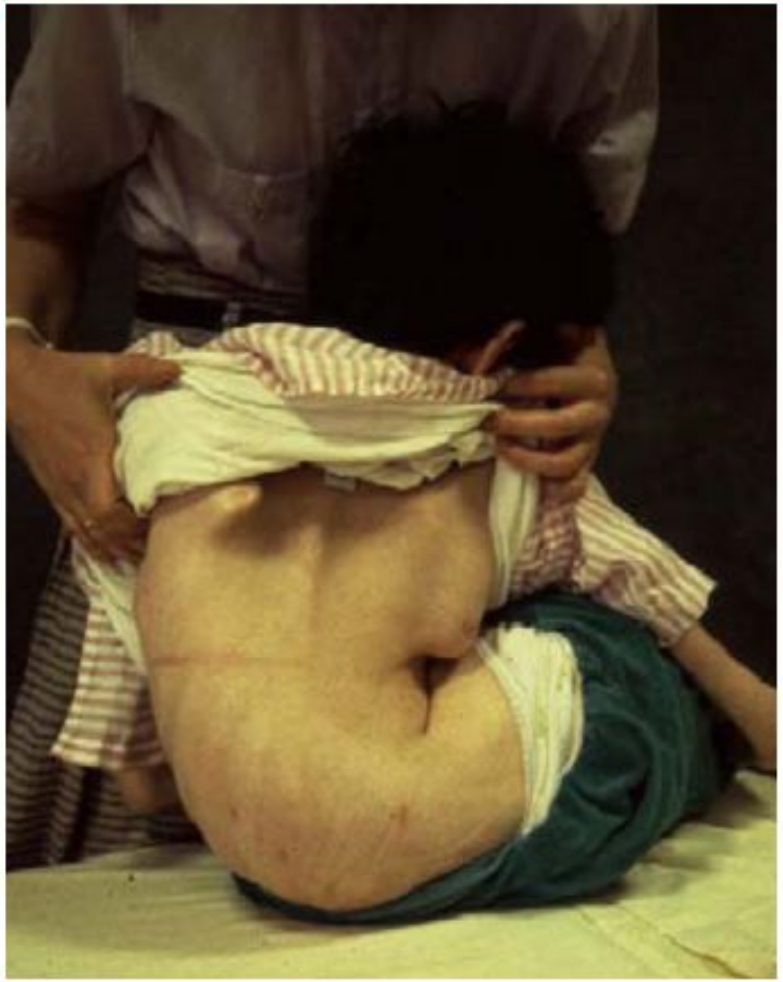
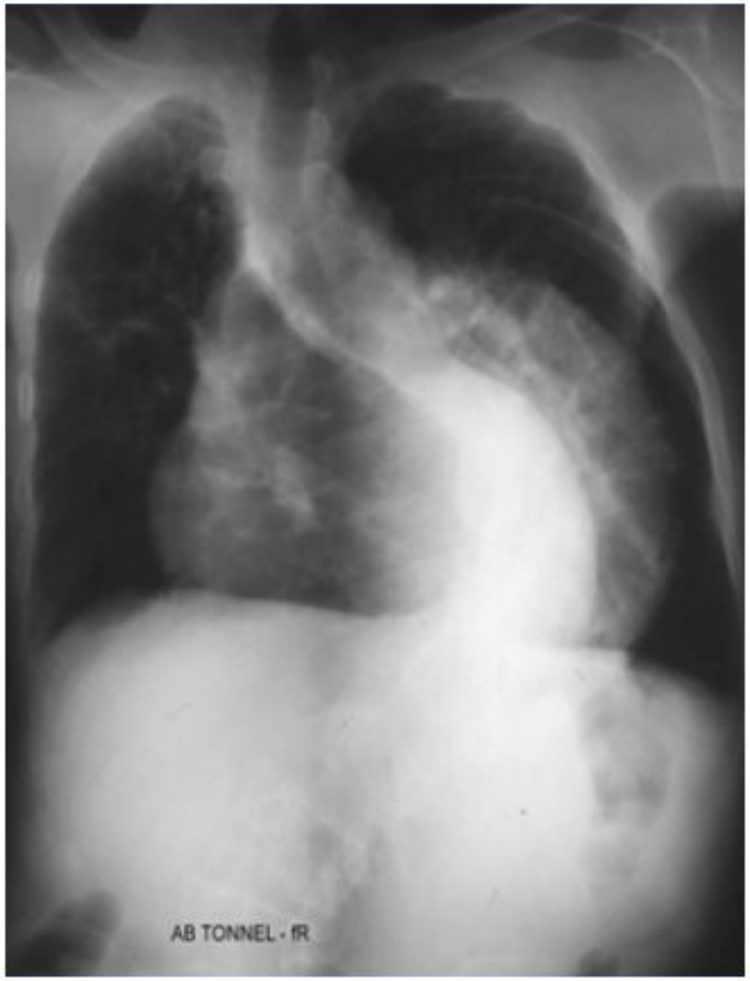


FIGURE 5. Schematic representation of pathologic links in neuromuscular disease. Inspiratory MW, inspiratory muscle weakness; Upper Airway MW, upper airway muscle weakness; Expiratory MW, expiratory muscle weakness; WOB, work of breathing.

Atteintes associées

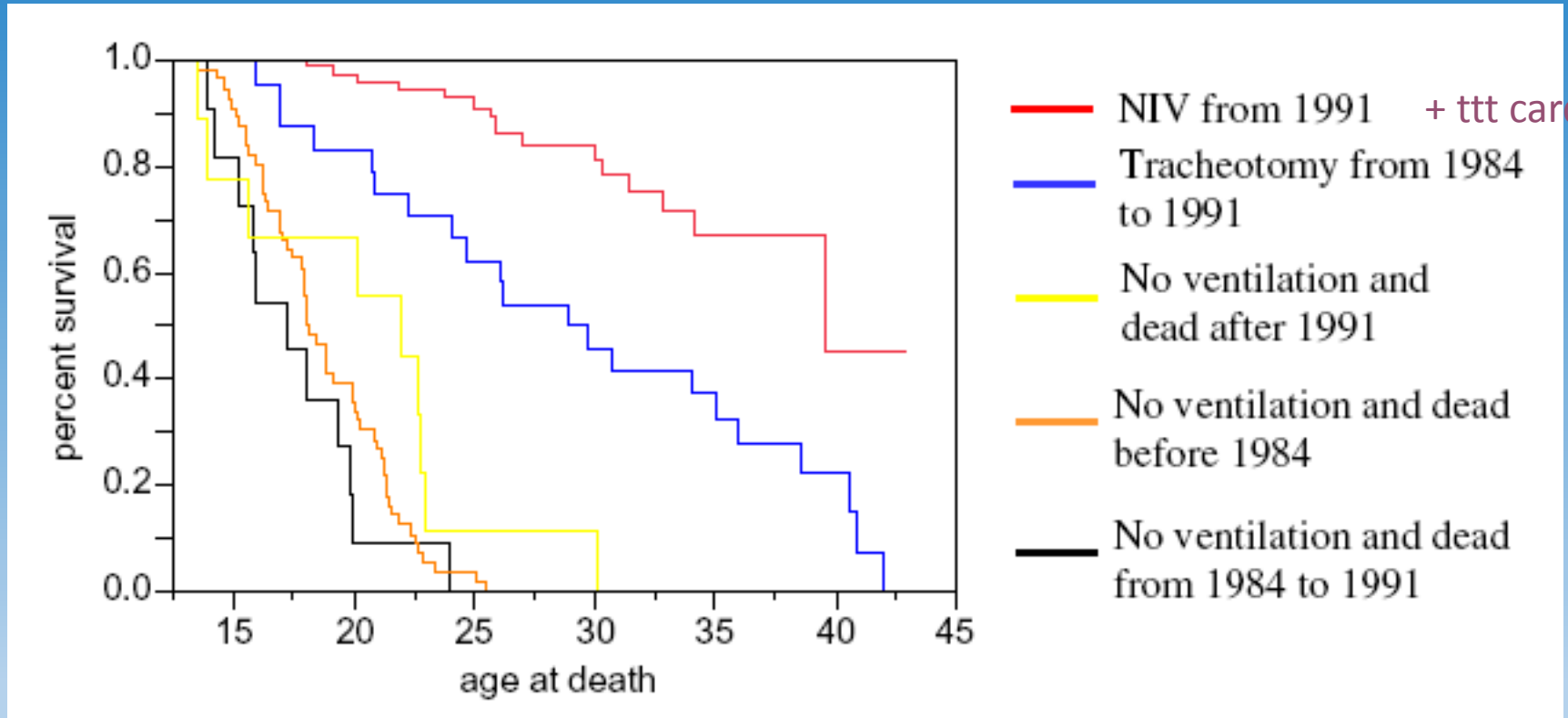
- Tb statique rachidienne: scoliose…aggrave l'atteinte respiratoire mais en ↓ (chir…)
- Atteinte cardiaque
- Tb du sommeil associé: SAS obstructif ou central et complexe (Steinert)




AB TONNEL - fR

VNI au long cours et maladies neuromusculaires

- **Peu d'études contrôlées** (SLA) car non éthiques compte tenu des études ouvertes et des essais négatifs de déventilation temporaire (Metha, *AJRCCM* 2001)
- La conduite d'essais cliniques contrôlés est recommandée pour préciser le bénéfice de la VNI dans l'IRC neuromusculaire ou pariétale et comparer les modes de ventilation (Annane, *Cochrane Database Review* 2007)



+ ttt cardio

Ishikawa *Neuromuscular disorders* 2011

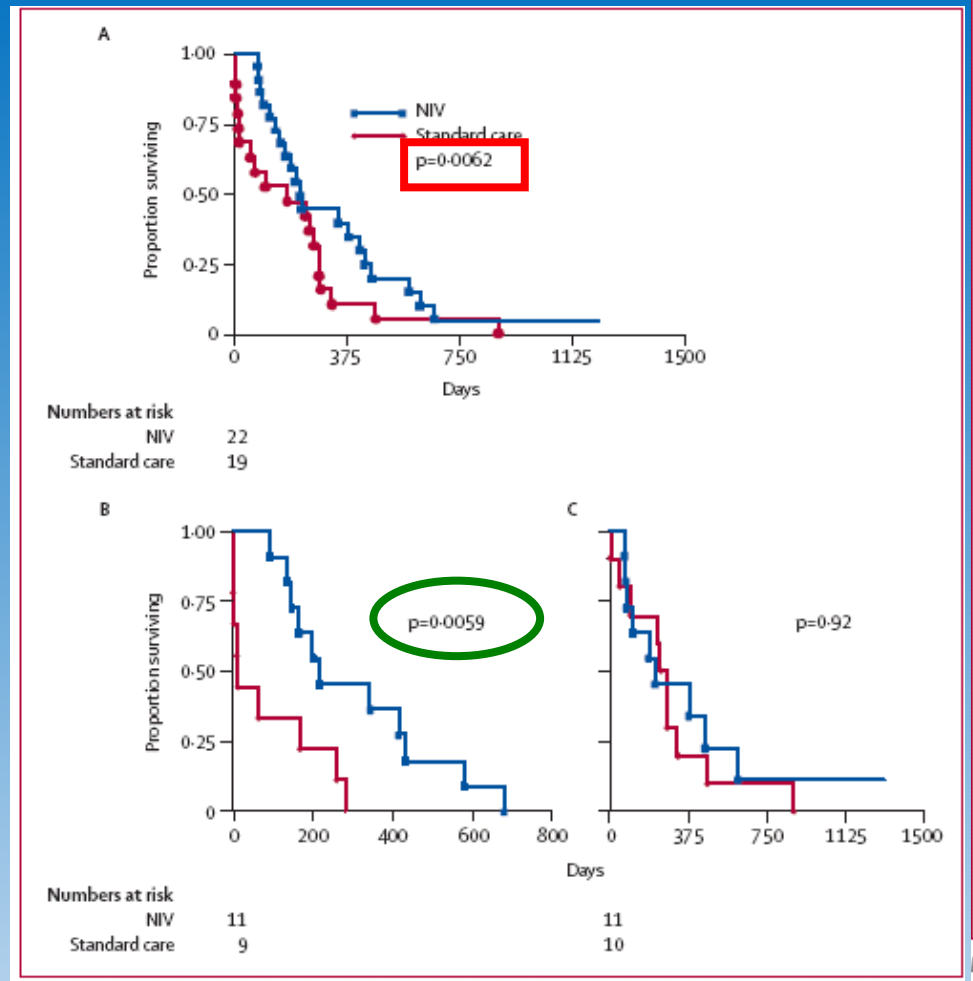


Figure 2: Survival from randomisation
A: all patients; B: patients with normal or moderately impaired bulbar function; C: patients with severe bulbar impairment.

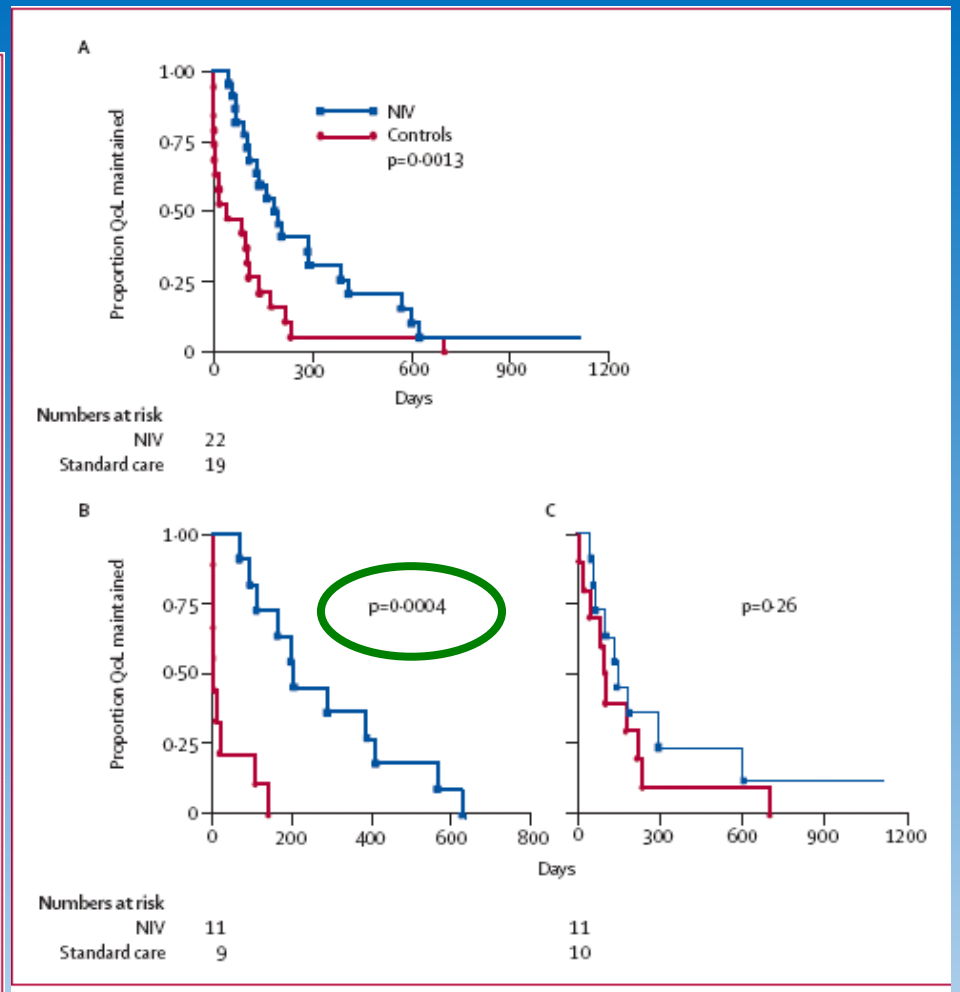


Figure 3: Time SAQLI symptoms domain maintained above 75% of prerandomisation assessment
A: all patients; B: patients with normal or moderately impaired bulbar function; C: patients with severe bulbar impairment. QoL=quality of life.

41 pt randomisés (orthopnée avec PI max < 60 et/ou Hcapnie symptomatique)

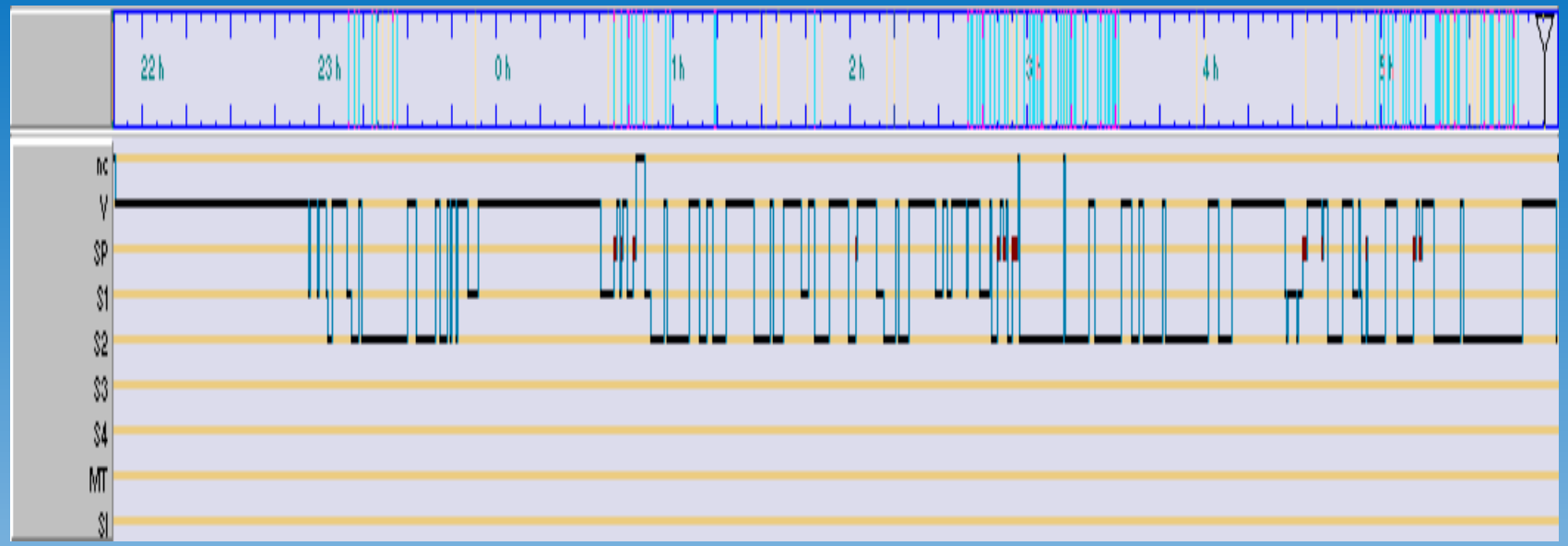
BIPAP ST

Bourke *Lancet Neurol* 2006

Bénéfices cliniques de la VNI à domicile

- Disparition en quelques semaines des signes d'hypoventilation : fatigue, somnolence diurne, céphalées matinales (Ellis, ARRD 1997)
- Corrections des troubles respiratoires du sommeil : apnées, désaturations en sommeil paradoxal (Ellis, ARRD 1997)

Mr S, 60 ans, SLA à forme spinale : Hypnogramme initial



Hypnogramme sous BiPAP, dès la nuit suivante

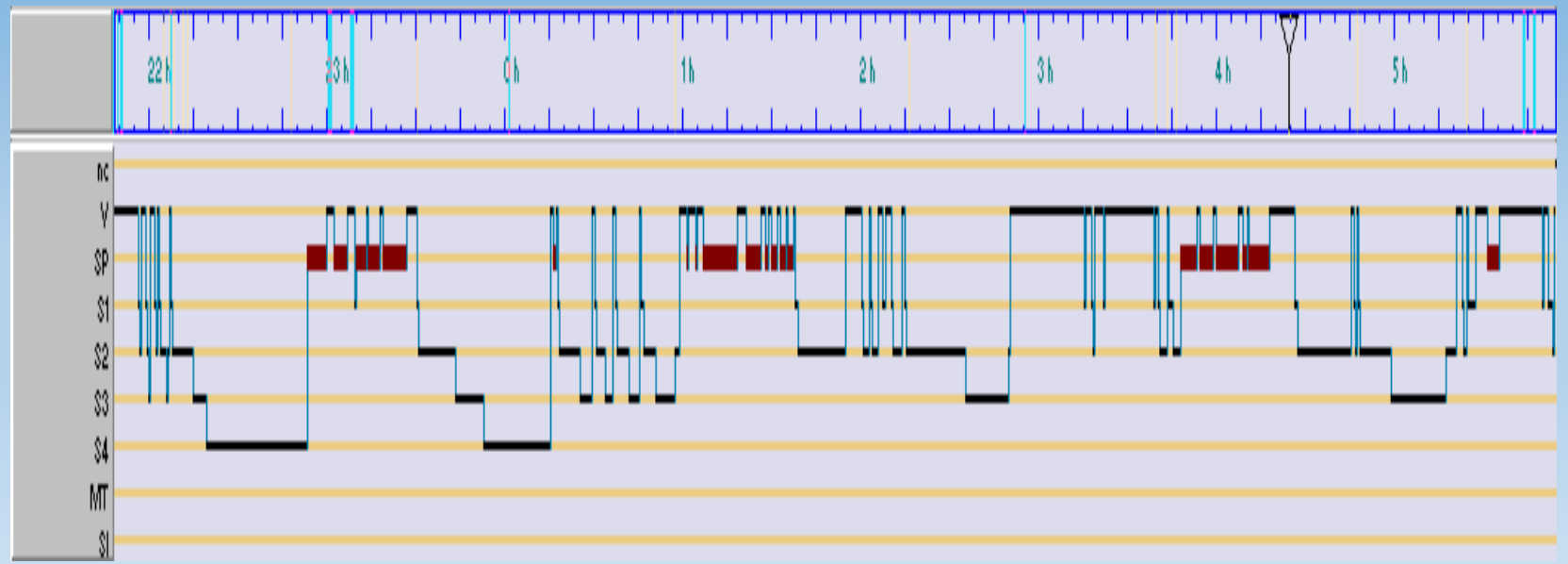
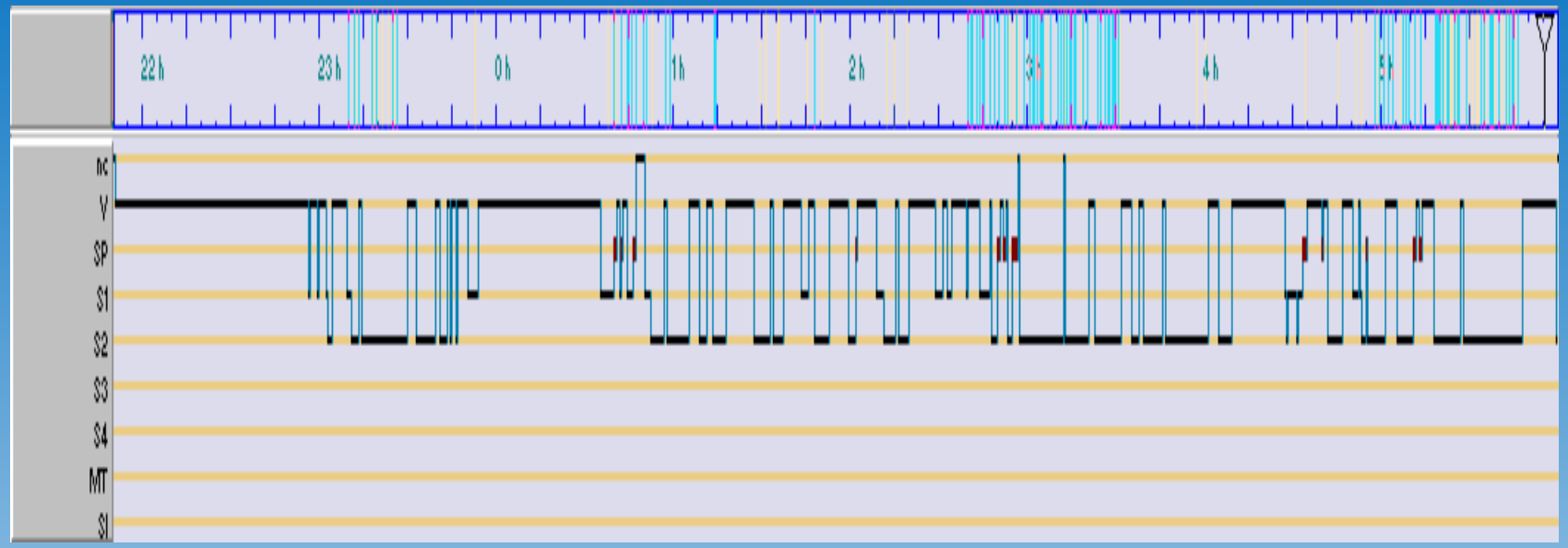


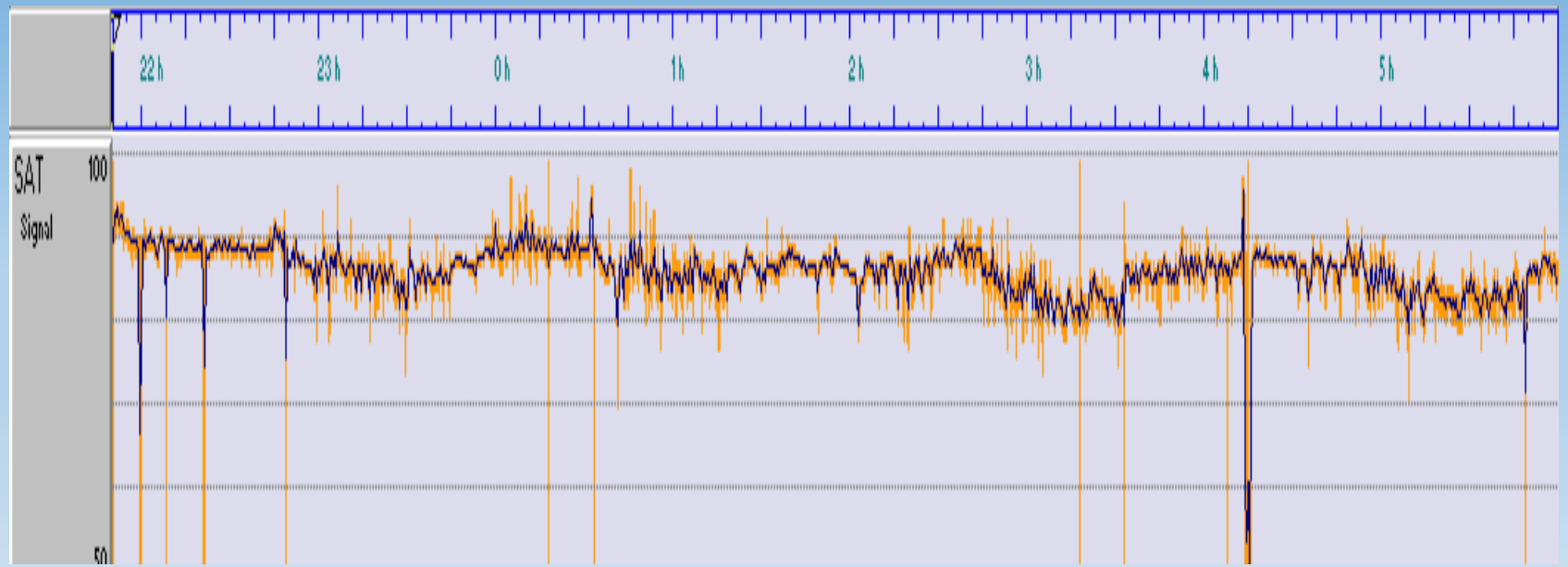
Table 3—*Polysomnographic Results at Baseline (Before NIPPV) and While Receiving NIPPV**

	Baseline	NIPPV
TST, min	272±45	375±30
RDI	22±6	1±1†
S1+S2, %	81±5	64±5
S3+S4, %	8±3	16±3†
REM, %	12±5	19±3
SE, %	59±8	83±5†
SaO_2<90%, min	160±53	8±4†
Mean SaO_2, %	88±3	95±1†
Minimal SaO_2, %	67±5	89±1†

Barbé *Chest* 1996

Mr S, 60 ans, SLA à forme spinale : Polysomnographie initiale





Mr S, 60 ans, SLA à forme spinale : Polysomnographie sous BiPAP

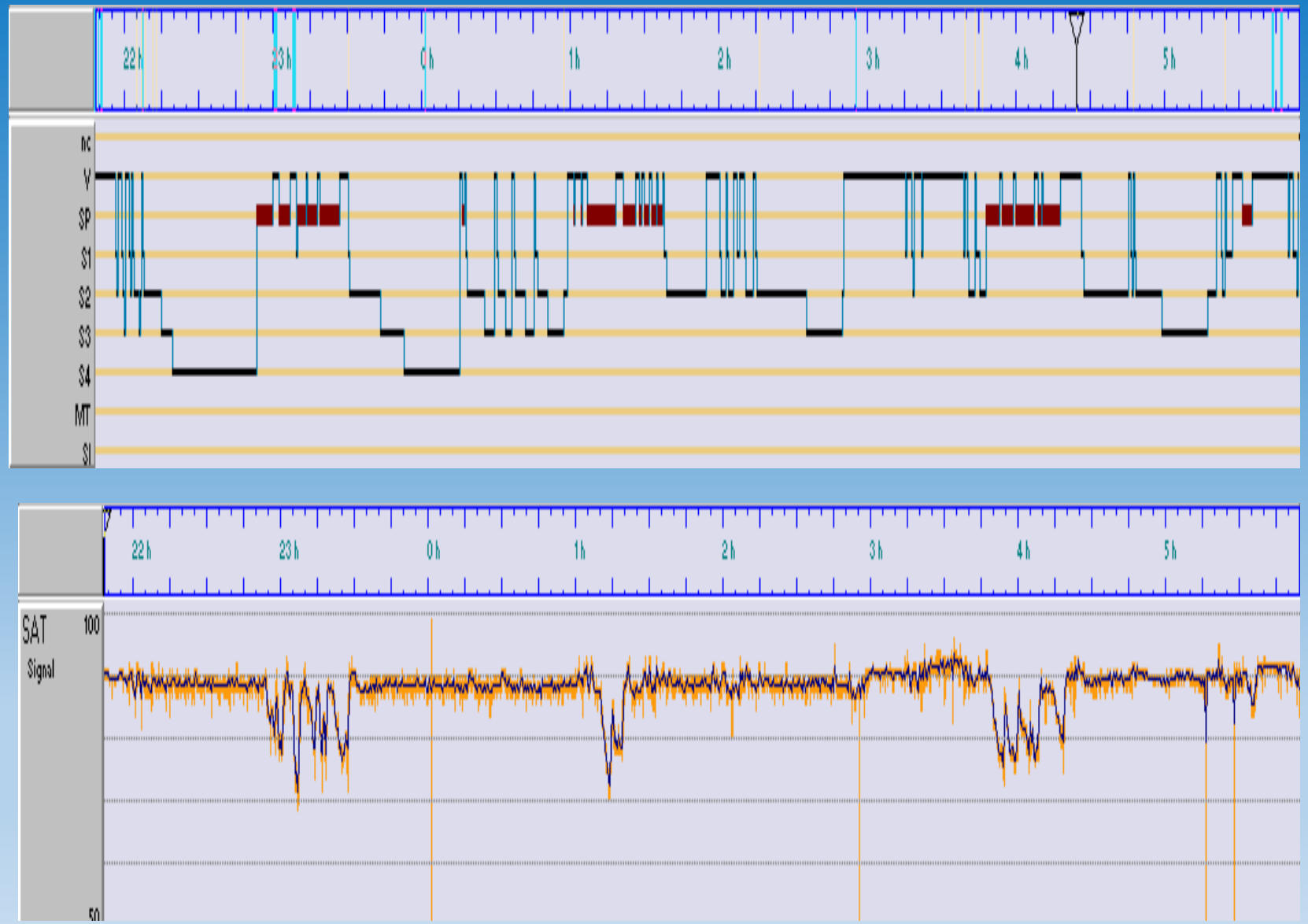


Table 2—Sleep structure in all, non-bulbar and bulbar patients before and after one month of NIV use.

	All (n = 22)		Non–bulbar (n = 13)		Bulbar (n = 9)	
	Pre	Post	Pre	Post	Pre	Post
TST (min)	317 (216–442)	351 (261–455)	311 (107–428)	364 (273–457)*	350 (287–454)	337 (241–436)
SE (%)	59 (40–72)	68 (50–80)	62 (19–81)	72 (52–85)*	59 (49–65)	68 (44–74)
N1 (%)	11 (6–32)	8 (4–14)*	23 (8–56)	5 (4–11)**	7 (5–13)#	9 (6–17)
N2 (%)	63 (37–75)	54 (46–63)	63 (36–77)	55 (49–63)	65 (42–75)	53 (33–64)
N3 (%)	1 (0–10)	15 (8–19)*	0 (0–6)	16 (13–19)*	7 (1–18)	12 (1–21)
REM (%)	9 (1–16)	18 (9–23)*	6 (0–12)	22 (10–23)**	17 (7–25)##	16 (9–23)
AAI (n/h)	37 (16–55)	17 (11–21)**	42 (36–82)	17 (12–26)**	16 (13–37)#	18 (11–20)

Vrijsen *J Clin Sleep Med* 2015

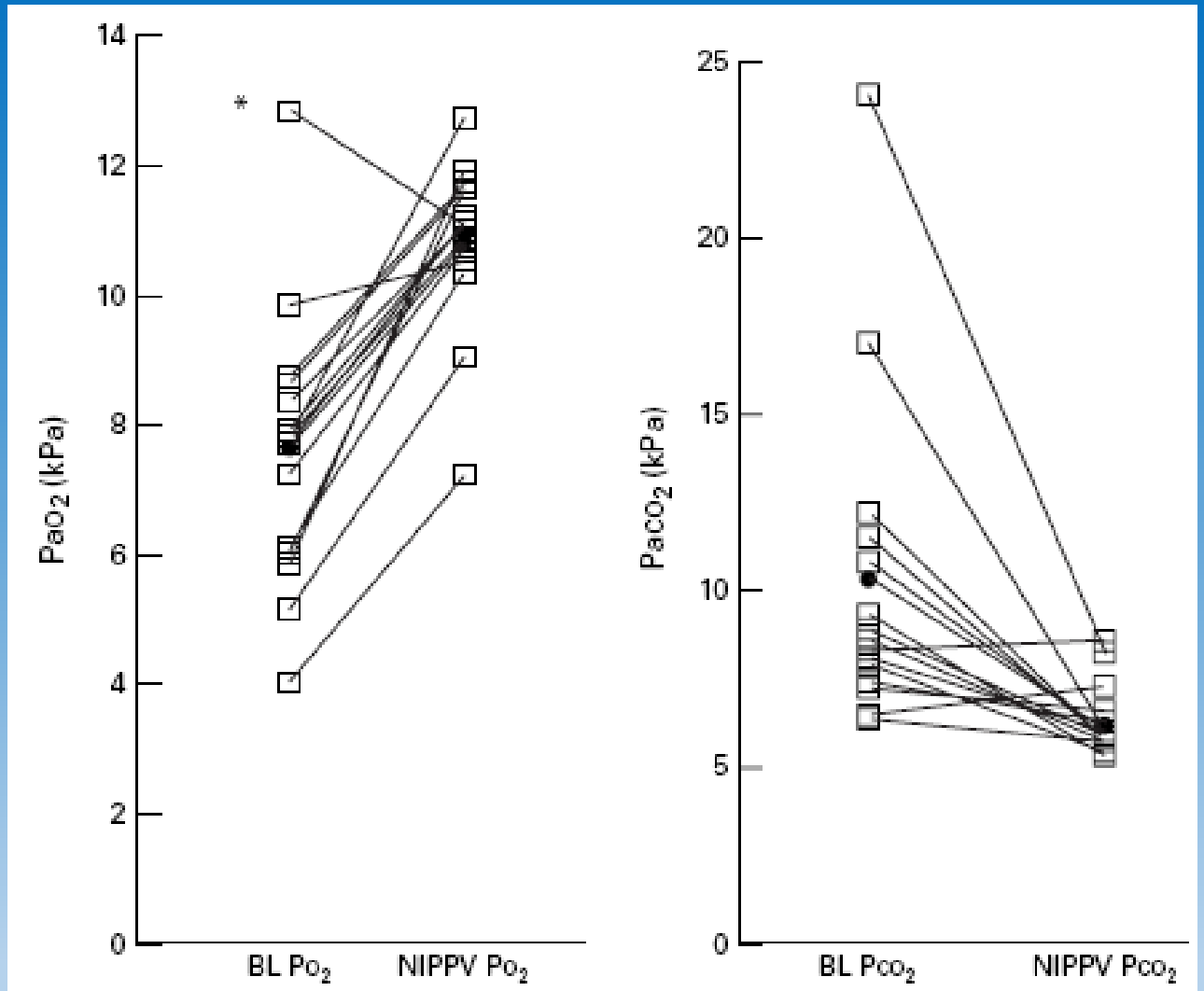


Figure 2 Effect of nasal intermittent positive pressure ventilation on arterial blood gas tensions in all patients. $BLPo_2$ = baseline arterial oxygen tension; $BLco_2$ = baseline arterial carbon dioxide tension; $NIPPVPo_2$ = arterial oxygen tension breathing spontaneously after starting NIPPV; $NIPPVPco_2$ = arterial carbon dioxide tension breathing spontaneously after starting NIPPV.

Simmonds *Thorax* 1998

Mechanisms of improvement of respiratory failure in patients with restrictive thoracic disease treated with non-invasive ventilation

Thorax 2005;60:754–760. doi: 10.1136/thx.2004.039388

A H Nickol, N Hart, N S Hopkinson, J Moxham, A Simonds, M I Polkey

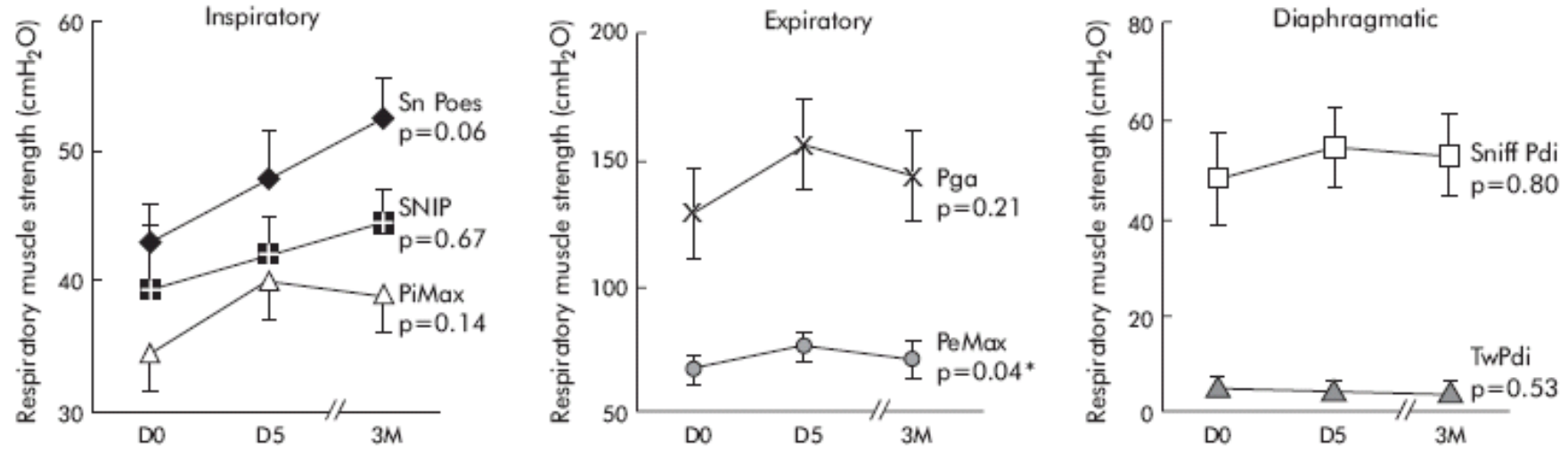


Figure 3 Effect of NIV on inspiratory, expiratory and diaphragmatic respiratory muscle strength. Mean (SE) values at day 0 (D0), day 5 (D5) and 3 months (3M) are shown. Sniff Poes, sniff oesophageal pressure; PiMax, maximum inspiratory pressure; SNIP, sniff nasal pressure; PeMax, maximum expiratory pressure; Pga, cough gastric pressure; Sniff Pdi, sniff transdiaphragmatic pressure; TwPdi, twitch transdiaphragmatic pressure. Significance values are given.

Table 3 Mean (SD) static lung and chest wall compliance (ml/cm H_2O) in the study patients

	Static lung compliance				Static chest wall compliance			
	D0	D5	3M	p value	D0	D5	3M	p value
All patients	80 (19)	103 (35)	96 (29)	0.20	90 (33)	97 (43)	98 (31)	0.98
Neuromuscular weakness	85 (18)	117 (32)	98 (31)	0.18	104 (29)	104 (41)	102 (30)	0.55
Kyphoscoliosis	71 (20)	72 (17)	91 (26)	0.12	63 (26)	80 (46)	90 (36)	0.25

Mechanisms of improvement of respiratory failure in patients with restrictive thoracic disease treated with non-invasive ventilation

A H Nickol, N Hart, N S Hopkinson, J Moxham, A Simonds, M I Polkey

Thorax 2005;60:754–760. doi: 10.1136/thx.2004.039388

Amélioration du drive respiratoire

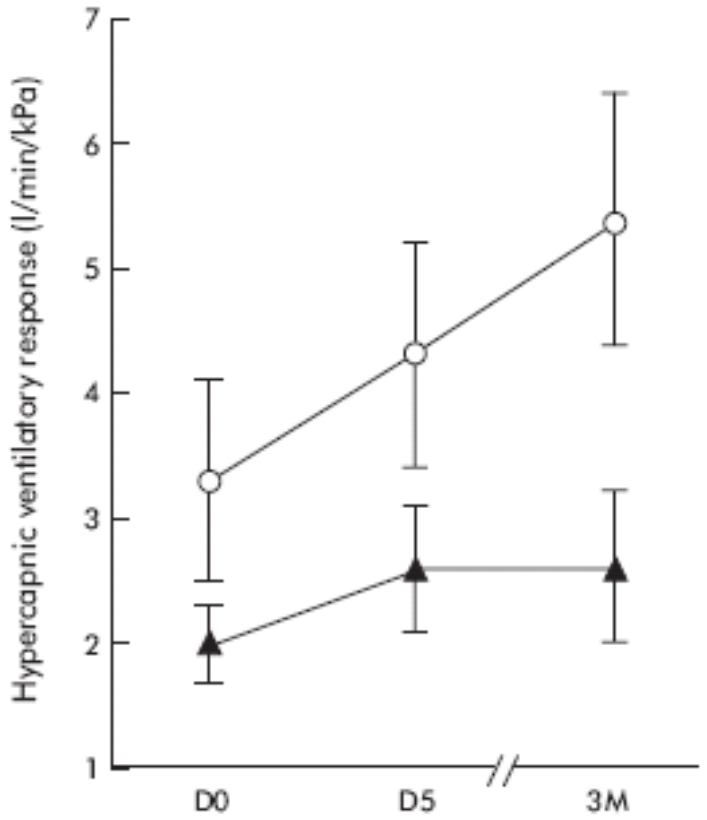


Figure 2 Effect of NIV on hypercapnic ventilatory response (HCVR) in patients with neuromuscular weakness (open circles) and kyphoscoliosis (closed triangles). Mean (SE) HCVR at day 0 (D0), day 5 (D5), and 3 months (3M) is shown. For the group as a whole there was a significant rise in HCVR ($p = 0.04$).

Indications de ventilation

Épisodes aigus

- Décompensation respiratoire aigue révélant la maladie ou d'une MNM sans évaluation respiratoire
- DRA chez patient NM connu et suivi

Indications de ventilation

Mise en route programmée

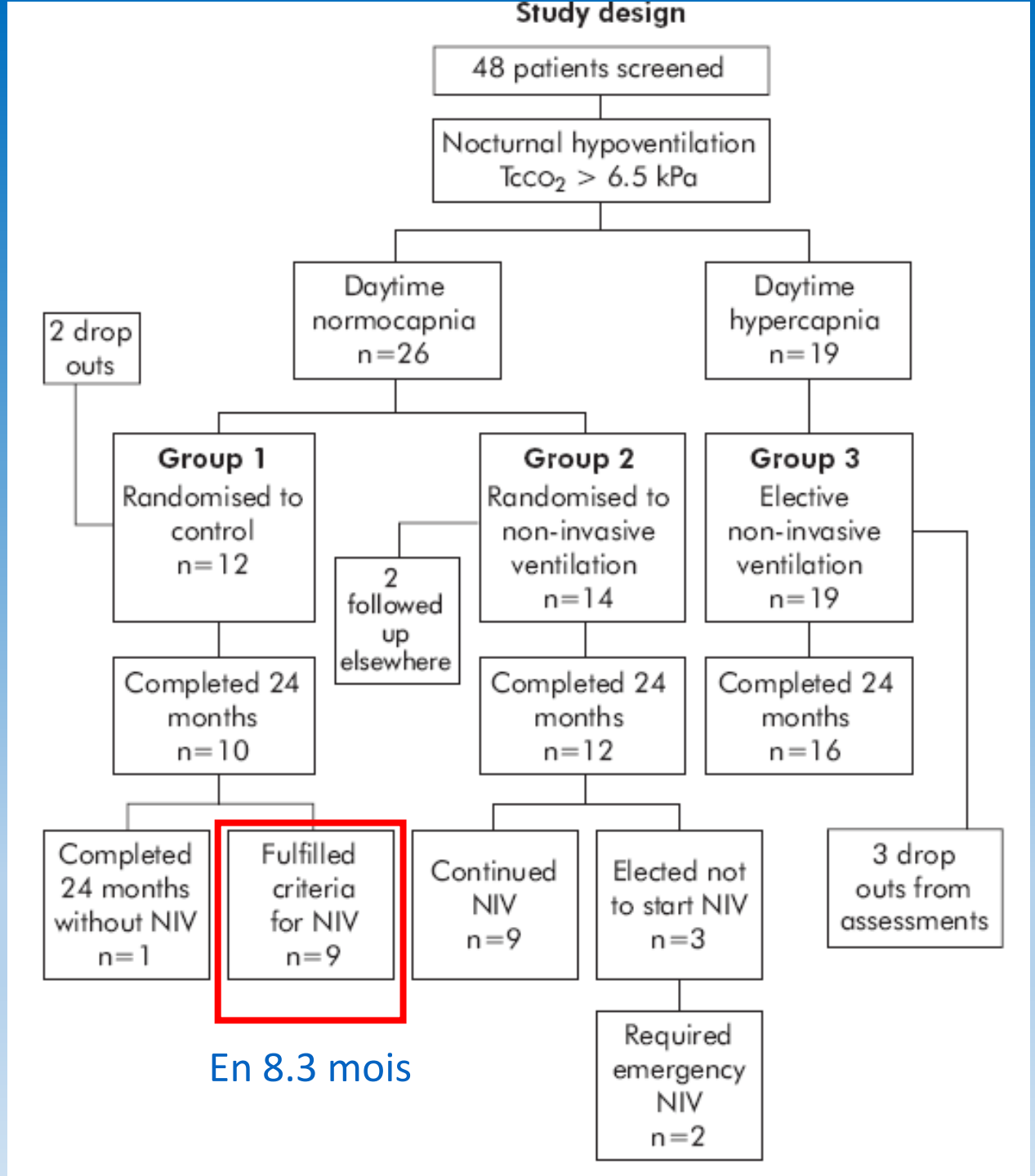
- Existence de **signes cliniques** (dyspnée, orthopnée, céphalées, asthénie, somnolence diurne, troubles du sommeil, amaigrissement, encombrement récidivant malgré aide à la toux…)
- Au moins **1** des **critères objectifs**

– paCO2 > 45 mmHg

– Désaturation nocturne < 88 % + de 5 minutes consécutives

– Hypercapnie nocturne ptcCO2 > 6,5 kPa pour pic

– CV ?



En 8.3 mois

Ward *Thorax* 2005

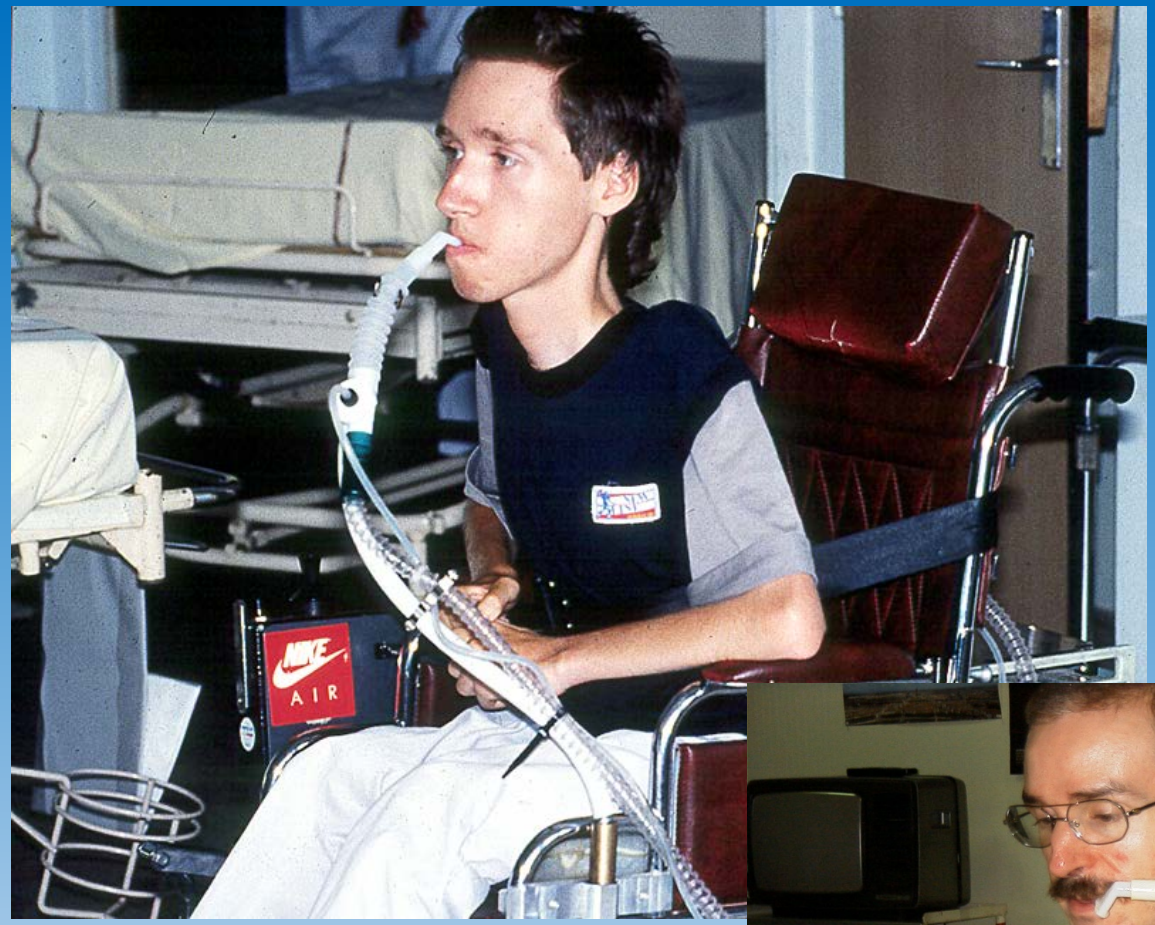
Table 1—*Possible Indications for NIV Support in NMD Patients*

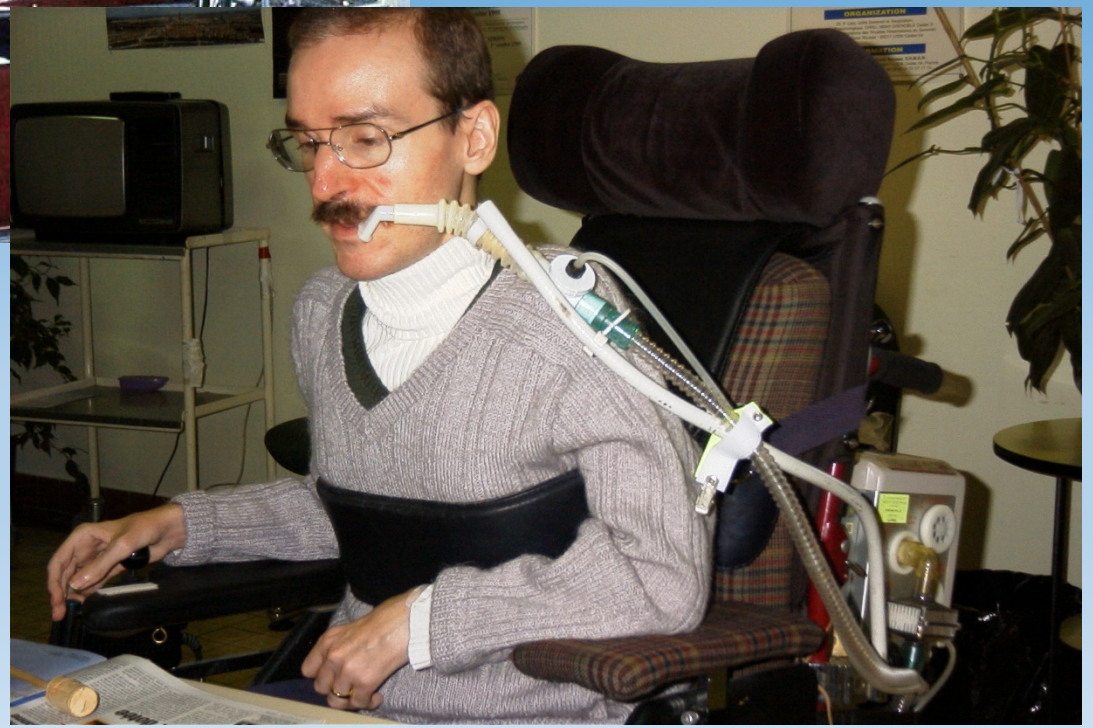
When to Start NIV?
Possibilities
To prevent respiratory decompensation
To alter chest wall/lung growth characteristics
During intercurrent chest infections
For perioperative period/gastric tube placement
During pregnancy
To rest respiratory muscles
To control nocturnal hypoventilation with or without symptoms
To treat established hypercapnic ventilatory failure
To palliate symptoms/end-of-life care

Simonds *Chest* 2006

Aspects pratiques en VNI

- D'abord nocturne. Diurne si pCO2 diurne > 45
- Mode ventilatoire préférentiel ?
- Subtilités ? Fréquence respiratoire
- Interfaces: nasal +++, masque moulé vs commerce ?, pipette, MNB en chronique ? ?
- Contrôle: au mieux polygraphie , GDS réveil, ± ptcCO2
- Objectifs en pratique:
  - Clinique: ↓ symptômes, bon sommeil, bonne tolérance
  - Paracliniques: normalisation SaO2 nocturne, et GDS nocturne et diurne.

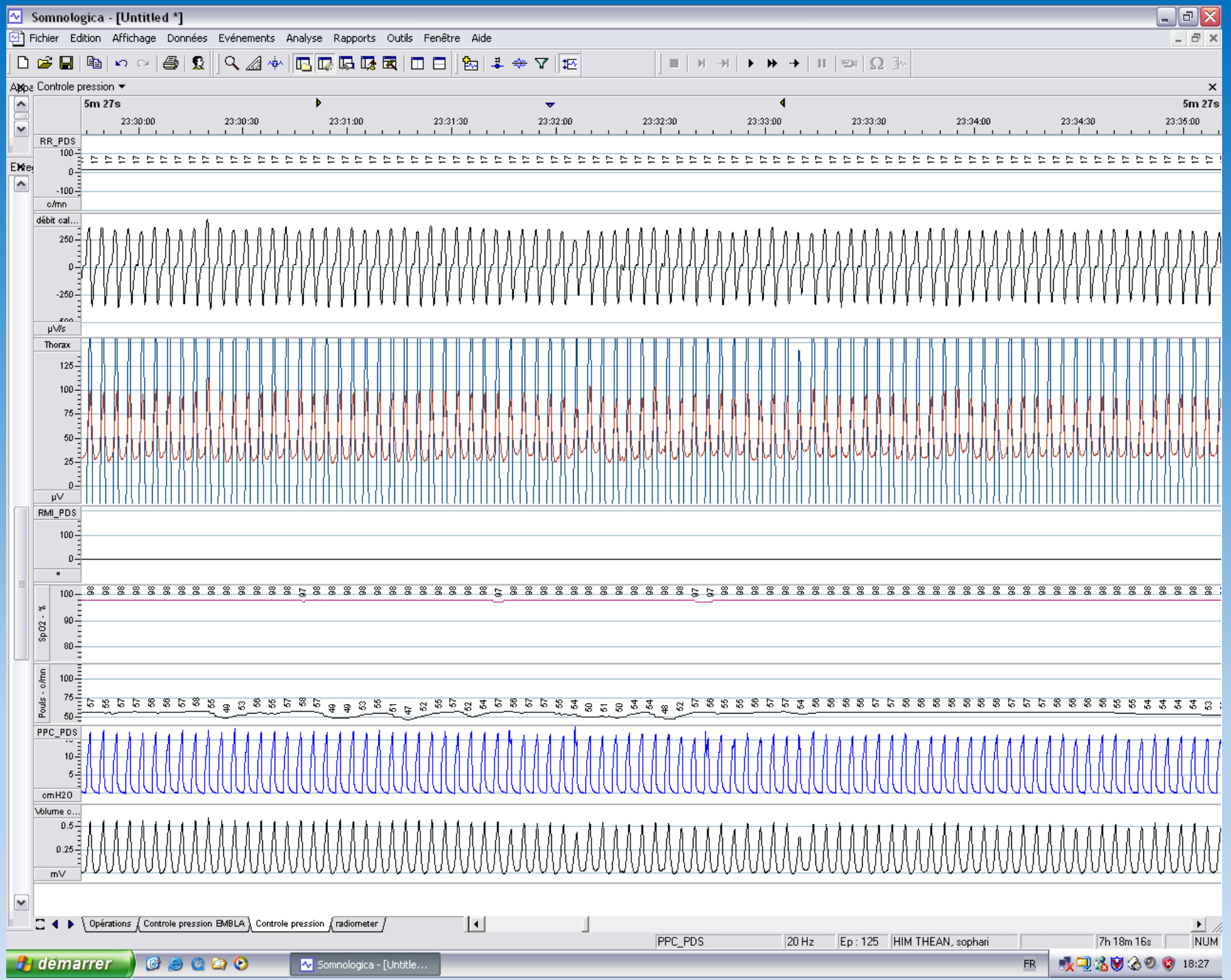




Ventilation à la pipette

Efficace

Toussaint *ERJ* 2006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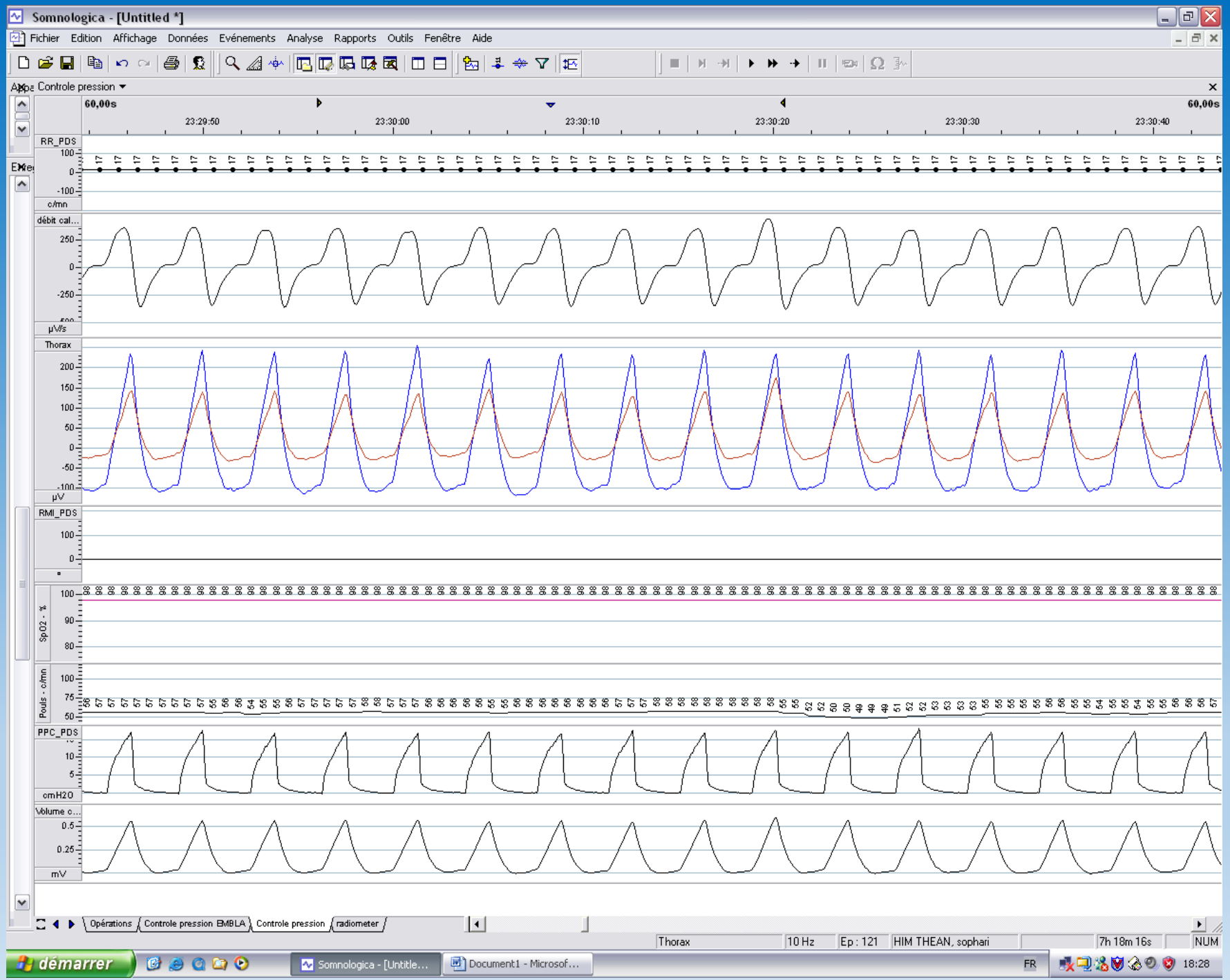
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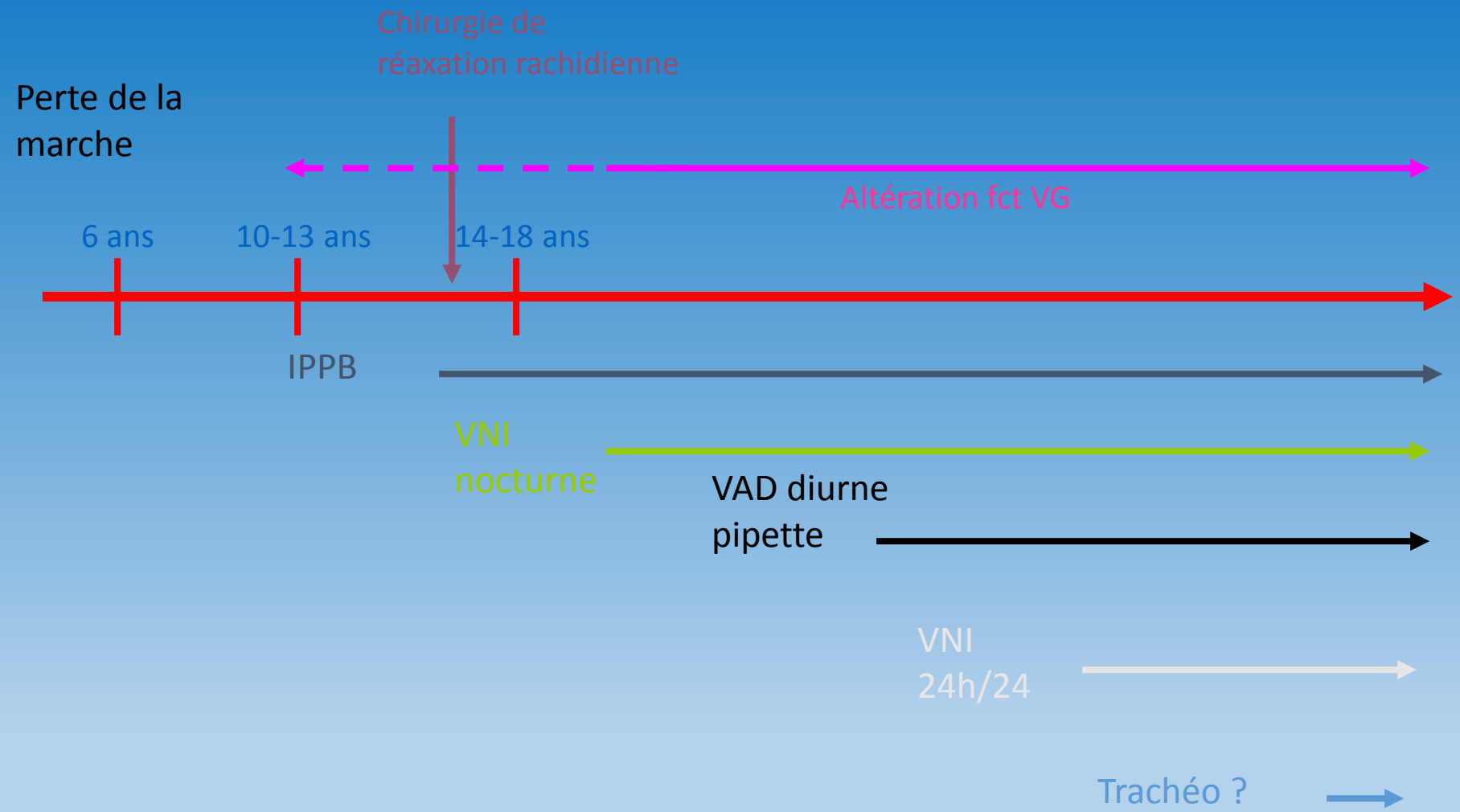
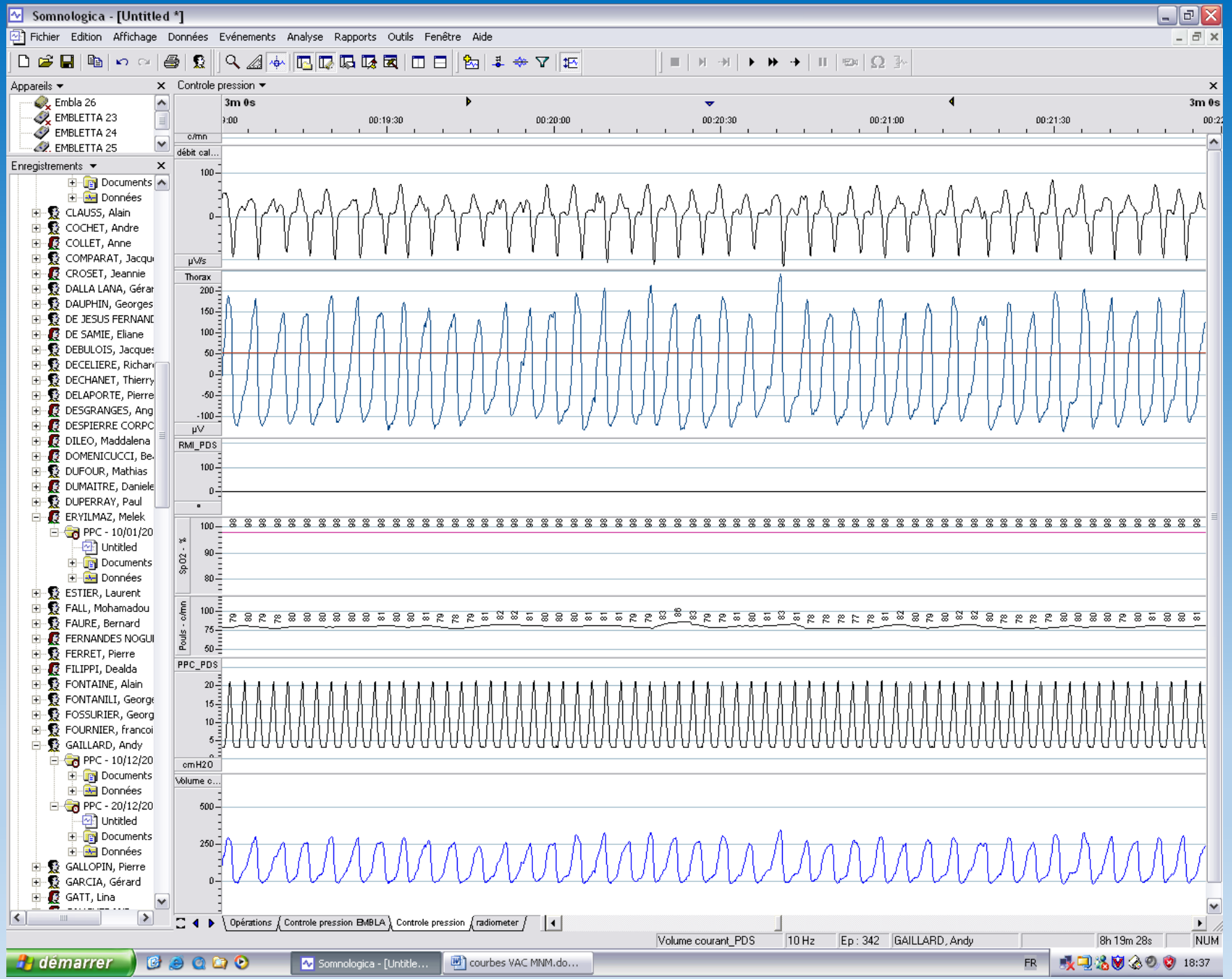
Évolution typique dans la myopathie de Duchenne

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DELAPORTE, Pierre
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SpO2 - %
Pouls - c/mn
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Air leaks during mechanical ventilation as a cause of persistent hypercapnia in neuromuscular disorders

	Nonhypercapnic patients (n=75)	Hypercapnic patients (n=20)	p (t test)
Age (years)	42±18	50±18	0.09
Height (cm)	170±52	164±12	0.6
Weight (kg)	54±24	68±22	0.02
Body mass index	19±7	25±7	0.001
Vital capacity (ml)	771±563	1050±649	0.07
Backup frequency (cycles/min)			
SB	20±2	20±2	0.4
MV	17±2	17±2	0.3
VI (l/min)	11.5±2.8	12.8±2.9	0.08
Period of ventilation (months)	50±89	27±27	0.25
Period of ventilation (h/day)	17±7	12±6	0.01
VTi (ml/kg)	13±6.3	12±3.8	0.3
VTe (ml/kg)	11.5±5.3	8.4±3.6	0.01
Air leaks (% of VTi)	19±14	32±22	0.003
pH			
SB	7.37±0.05	7.37±0.03	0.06
MV	7.45±0.05	7.38±0.02	<0.0001
$PaCO_2$ (mmHg)			
SB	46.5±6.0	51.8±6.0	0.0006
MV	35.3±6.0	50.3±4.5	<0.0001

Gonzalez, *ICM* 2003

Air leaks during mechanical ventilation as a cause of persistent hypercapnia in neuromuscular disorders

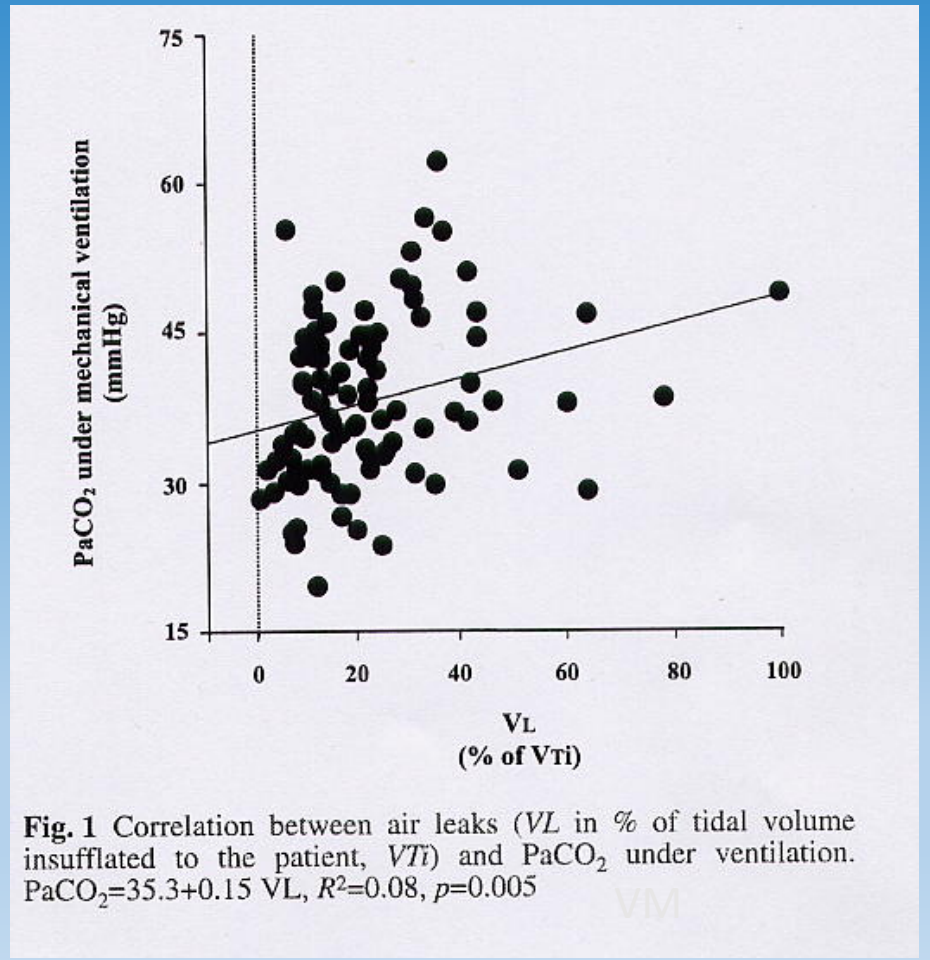


Fig. 1 Correlation between air leaks (*VL* in % of tidal volume insufflated to the patient, *VTi*) and $PaCO_2$ under ventilation. $PaCO_2$=35.3+0.15 VL, R^2=0.08, p=0.005

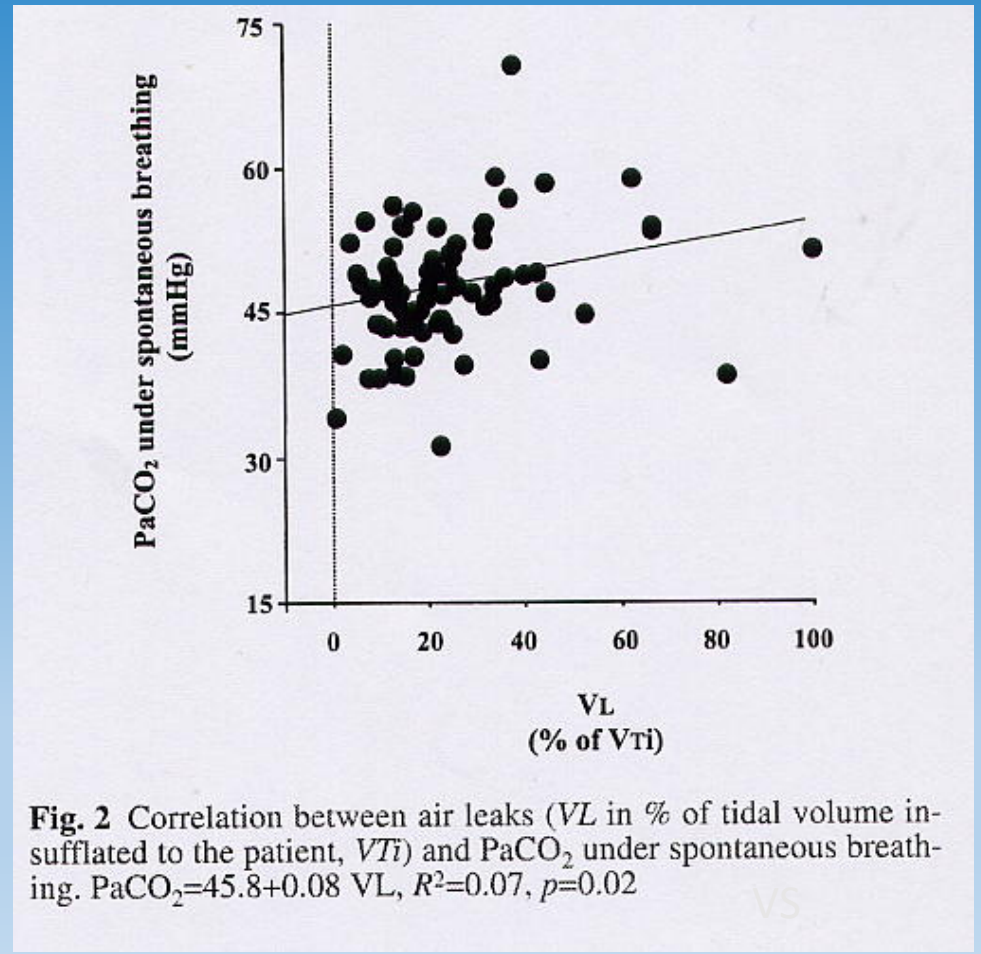
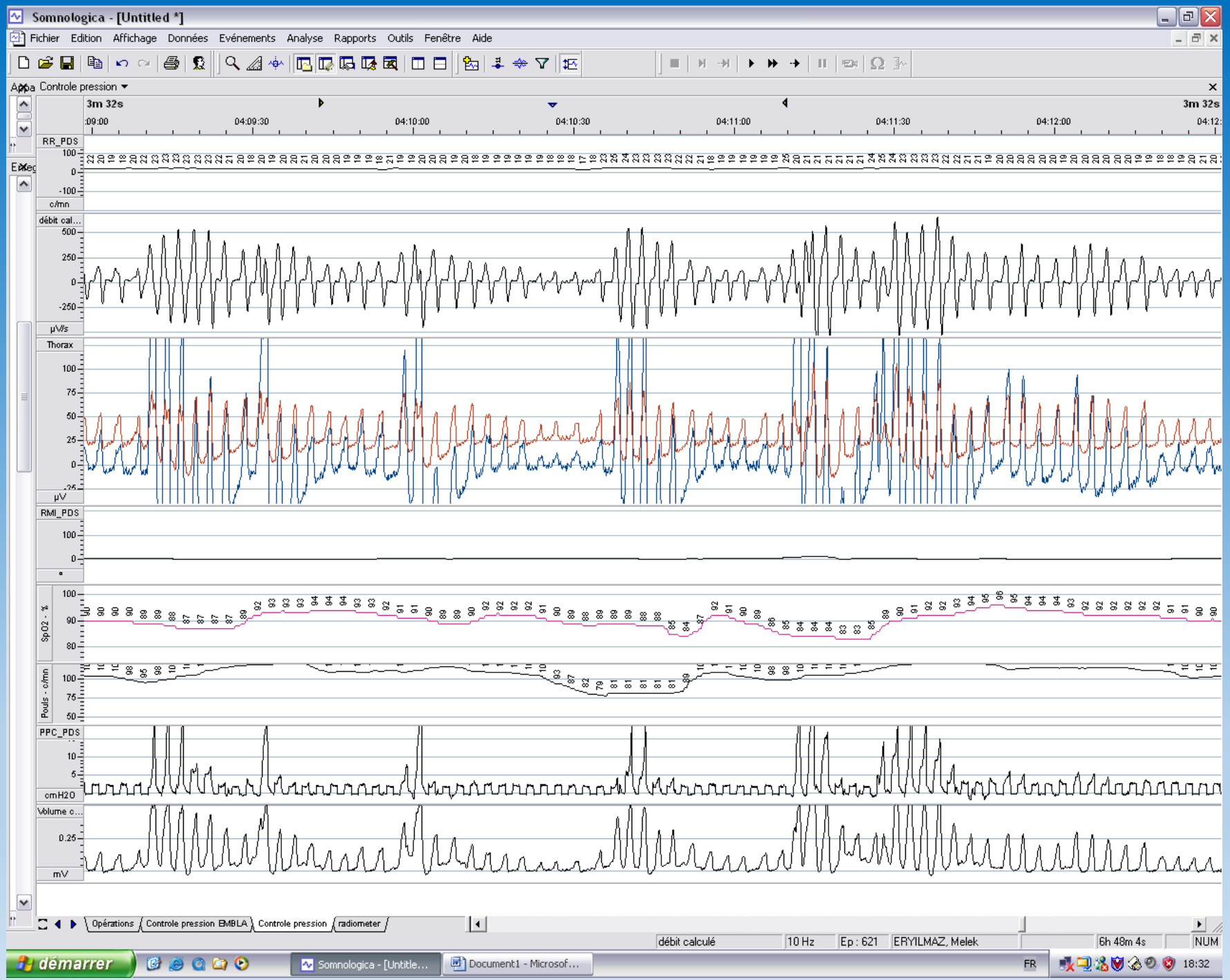


Fig. 2 Correlation between air leaks (*VL* in % of tidal volume insufflated to the patient, *VTi*) and $PaCO_2$ under spontaneous breathing. $PaCO_2$=45.8+0.08 VL, R^2=0.07, p=0.02

Gonzalez, *ICM* 2003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µV
RMI_PDS
SpO2 - %
Pouls - c/mn
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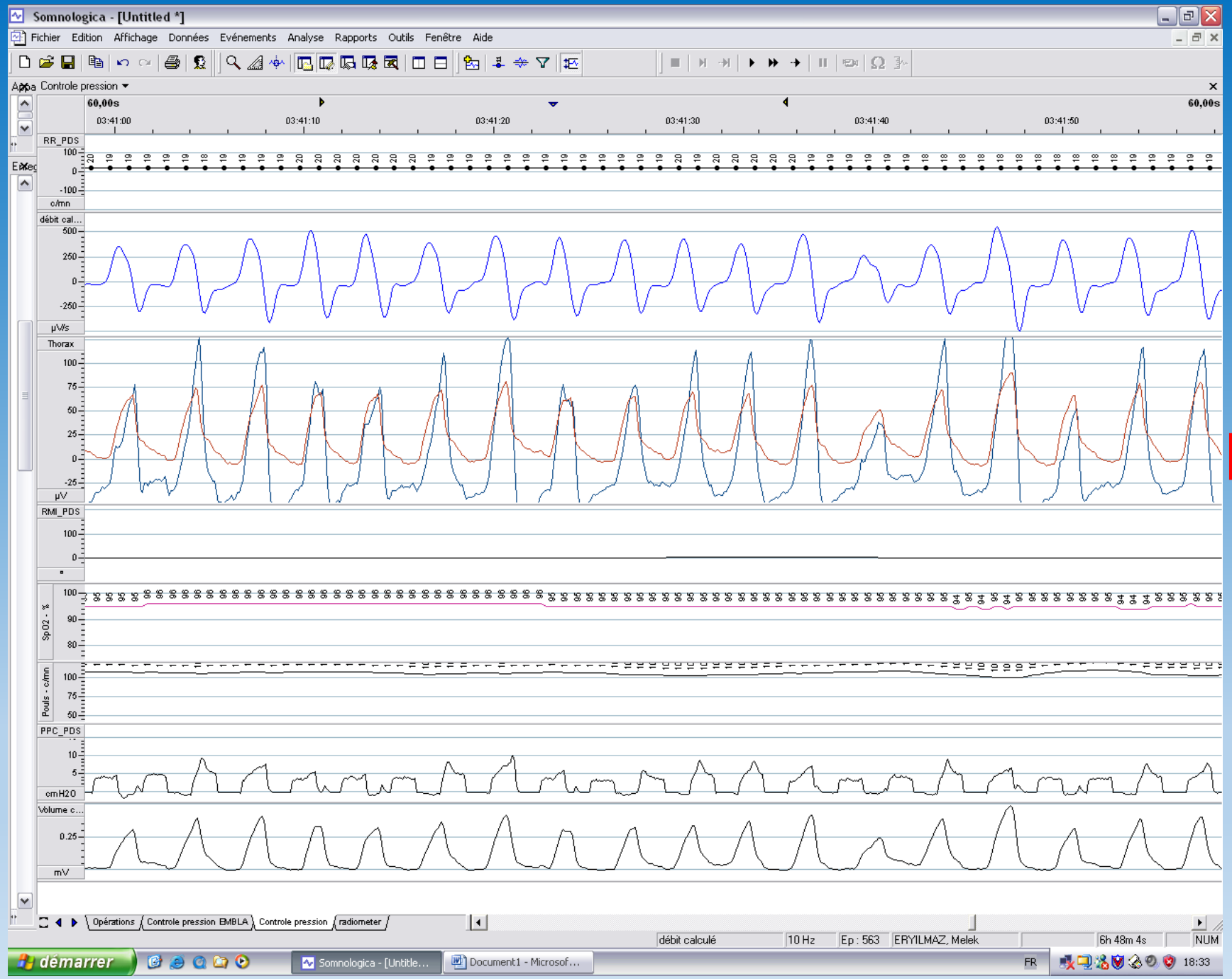
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µV/s
Thorax
µV
RMI_PDS
SpO2 - %
Pouls - c/mn
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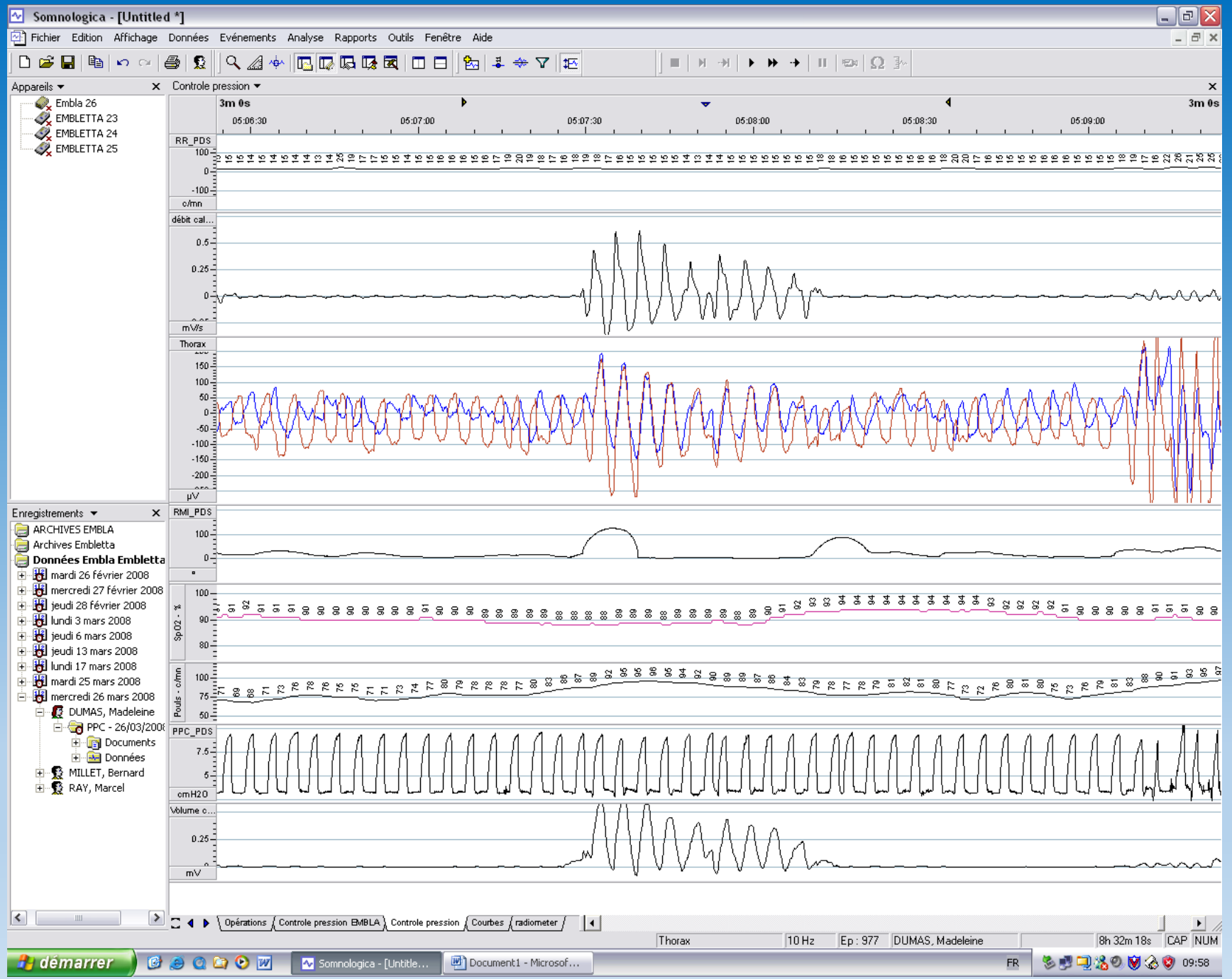
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RMI_PDS
Sp02 - %
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cmH2O
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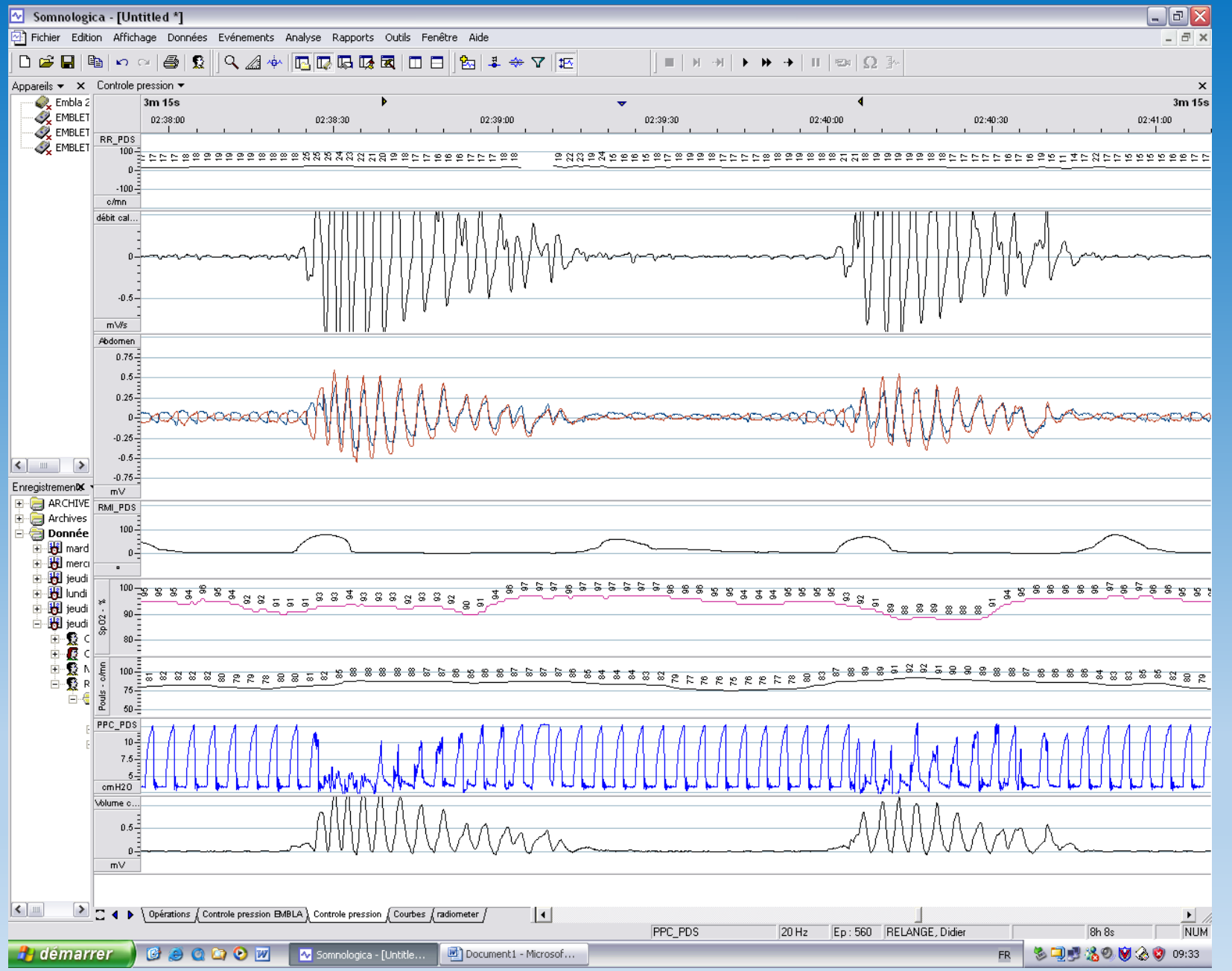
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mV
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SpO2 - %
Pouls - c/mn
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Les limites

- Connaissance
- Évaluation encombrement: difficile
- Matériel disponible (ventilation et kiné)
- Le patient lui-même:

- dépendance ventilatoire
- Efficacité de la technique de désencombrement (pas reproductible)
- Efficacité de la ventilation

En chronique ≠ en aigu

***Table 2** Factors influencing NIPPV tolerance (multivariate logistic regression)*

Variable	Odds ratio (95% CI)	*p* Value
Age at NIPPV	1.01 (0.97–1.04)	0.649
ALS-FRS score	1.04 (0.96–1.12)	0.290
Mild/moderate bulbar symptoms	6.09 (1.18–31.52)	0.031*
PEG placement	0.52 (0.18–1.55)	0.092

* Significant ($p < 0.05$)

NIPPV = noninvasive positive-pressure ventilation; ALS-FRS = ALS Functional Rating Scale; PEG = percutaneous endoscopic gastrostomy.

***Table 3** Multivariate analysis (Cox model) showing independent predictors of survival after initiation of NIPPV in the overall group of patients*

Variable	Risk ratio (95% CI)	*p* Value
NIPPV use ≥4 h/day	0.32 (0.13–0.78)	0.013*
FVC% slope after NIPPV	0.78 (0.65–0.94)	0.010*

* Significant ($p < 0.05$).

NIPPV = noninvasive positive-pressure ventilation; FVC% = forced vital capacity (percentage of predicted).

71 pt:

- 44 tolérants VNI
- 27 non tolérants
- BIPAP ST

Lo Cocco *Neurology* 2006

Oral appliance to assist non-invasive ventilation in a patient with amyotrophic lateral sclerosis

?

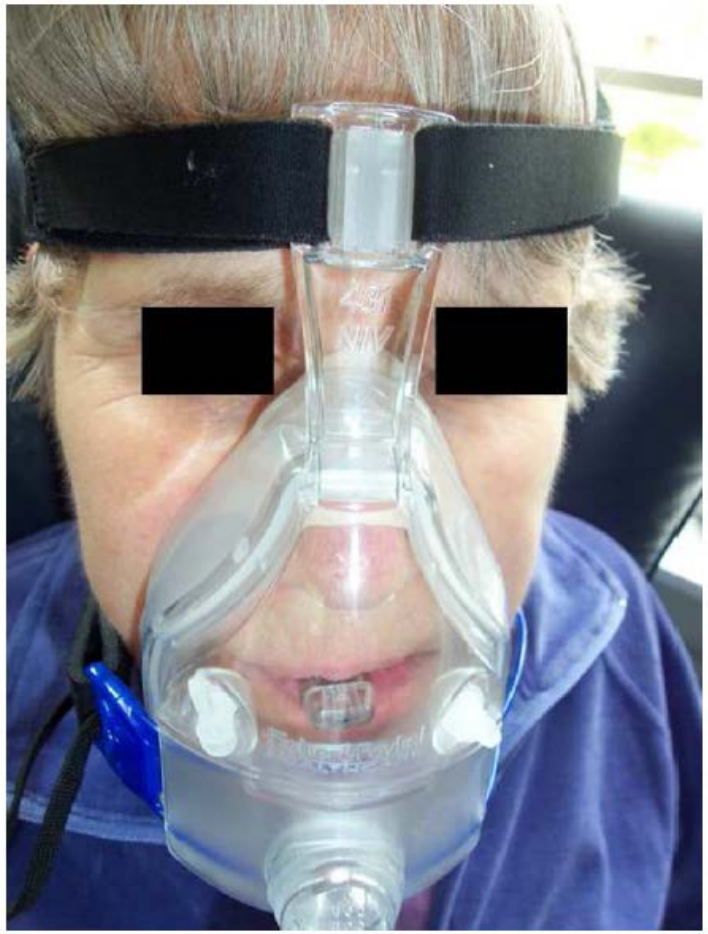

Fig. 1 A combination of a mandibular advancement device and an oronasal mask

Veldhuis *Sleep breath* 2015

Complications respiratoires et particulièrement encombrement des VAS, facilitée par une toux inefficace: **principale cause de mortalité** et morbidité dans la SLA

Table 3 Most frequent diagnoses and procedures in hospitalized patients with ALS

Diagnosis/procedure	Primary diagnosis, n (%)*	Total occurrence, n (%)†
Diagnoses		
Acute respiratory failure	134 (8.4)	359 (22.4)
Pneumonitis from aspiration of food particles	133 (8.3)	201 (12.6)
Pneumonia, organism unspecified	85 (5.3)	150 (9.4)
Pneumonia, *Pseudomonas*	29 (1.8)	67 (4.2)
Volume depletion	25 (1.6)	212 (13.2)
Procedures‡		
PEG	136 (8.5)	211 (13.3)
Mechanical ventilation >96 h	82 (5.1)	182 (11.4)
Temporary tracheostomy	69 (4.3)	77 (4.8)
Mechanical ventilation <96 h	55 (3.4)	84 (5.2)
Permanent tracheostomy	38 (2.4)	40 (2.5)

40 / 53 pt : infection respiratoire dans l'année
Sancho *AJRCCM* 2007

Lechtzin *Neurology* 2001

Alternatives to Endotracheal Intubation for Patients with Neuromuscular Diseases

Servera E, Sancho J, Zafra MJ, Catalá A, Vergara P, Marin J: Alternatives to endotracheal intubation for patients with neuromuscular diseases. Am J Phys Med Rehabil 2005;84:851–857.

- 17 patients (11 SLA + 4 Duchenne)
- prospective
- En IRA
- VNI en VAC, continue (pipette + nasal) et In-Exsufflation
- Atteinte bulbaire: mauvais PNT
- Bons objectifs pour contrôle hématose

Alternatives to Endotracheal Intubation for Patients with Neuromuscular Diseases

Servera E, Sancho J, Zafra MJ, Catalá A, Vergara P, Marín J: Alternatives to endotracheal intubation for patients with neuromuscular diseases. Am J Phys Med Rehabil 2005;84:851–857.

- Succès = 79,2% (ni IOT ni décès)
- Échec: 20,8 % (5 SLA dont 1 arrêt cardiaque, 1 palliatif)
- Si comparaison autres études: ↓ mortalité et ↓ séjour hospitalier

Extubation of Patients With Neuromuscular Weakness : A New Management Paradigm

John Robert Bach, Miguel R. Gonçalves, Irram Hamdani and Joao Carlos Winck

Chest 2010

113 pt New J
44 pt Portugal

Exclusion SLA bulbaire

Table 2—*Demographic Data*

Characteristics	Group 1[a]	Group 2[b]	Group 3[c]	Total
Subjects, No. (%)	74 (47)	45 (29)	38 (24)	157 (100)
Sex, No. (%)	52 male (70)	28 male (62)	17 male (45)	97 male (62)
	22 female (30)	17 female (38)	21 female (55)	60 female (38)
Diagnoses, No. (%)	ICUMy, 15 (20)	SMA, 10 (22)	SMA, 10 (26)	SMA, 25 (16)
	SCI, 13 (18)	MD, 9 (20)	DMD, 9 (24)	MD, 22 (14)
	ALS, 11 (15)	DMD, 8 (18)	MD, 5 (13)	DMD, 20 (13)
	MG, 9 (12)	MG, 6 (13)	PPS, 5 (13)	SCI, 17 (11)
	MD, 8 (11)	PPS, 4 (9)	oNMD, 4 (11)	ALS, 16(10)
	oNMD, 8 (11)	oNMD, 4 (9)	SCI, 3 (8)	oNMD, 16 (10)
	SMA, 5 (7)	ALS, 3 (7)	ALS, 2 (5)	ICUMy, 15 (10)
	DMD, 3 (4)	SCI, 1 (2)	. . .	MG, 15 (10)
	PPS, 2 (3)	. . .	. . .	PPS. 11 (7)
Use of NIV preintubation, No. (%)	No NIV, 51 (69)	No NIV, 24 (53)	No NIV, 21 (55)	No NIV, 96 (61)
	Cont, 10 (14)	Cont, 7 (16)	Cont, 3 (8)	Cont, 20 (13)
	Noct, 13 (17)	Noct, 14 (31)	Noct, 14 (37)	Noct, 41 (26)

ALS = amyotrophic lateral sclerosis; Cont = continuous noninvasive ventilation; DMD = Duchenne muscular dystrophy; ICUMy = ICU-acquired neuromuscular disease; MD = muscular dystrophy; MG = myasthenia gravis; NIV = noninvasive ventilation; Noct = nocturnal noninvasive ventilation; oNMD = other neuromuscular disease; PPS = postpolio syndrome; SCI = spinal cord injury; SMA = spinal muscular atrophy, including types 1, 2, and 3, and other neuromuscular disease.

[a]Local patients.

[b]Patients transferred after failing extubations in other institutions.

[c]Patients transferred after failing multiple spontaneous breathing trials in other institutions.

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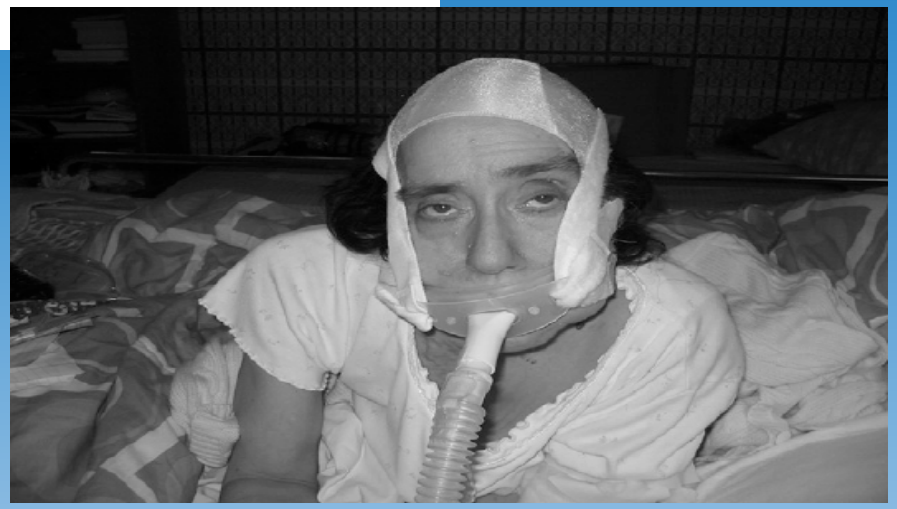
Chest 2010



VNI continue + MAC (tout intervenant)

172 extubations pour 157 pt

-PCF > 160 l/min: 98 succès (100 %)

-PCF < 160 l/min: 59 sur 74 (80 %) = succès dont 52 /60 dès 1er essai

-1 SLA et 1 FSH trachéo

-131/ 157 vivants

-Connaissance VNI + MAC avant IOT facilite sevrage

Quand proposer la trachéotomie ?

- VAD inefficace: hypercapnie sévère sous VNI (fuites buccales, pipette inefficace…), dysfonction glottique
- Troubles de déglutition sévères et compliqués
- Épisodes de décompensations respiratoires aigus récurrents ?
- Dépendance ventilatoire totale et intolérance interface (lésions cutanées…) ?
- Décision anticipée et réfléchie

Ventilation par trachéotomie

- Permet une ventilation assistée totale, avec un circuit étanche
- La canule de trachéotomie protège l'arbre respiratoire de l'encombrement salivaire et permet l'aspiration directe des sécrétions
- Permet aussi une ventilation ballonnet dégonflé