VNI et Mucoviscidose : Quelles indications ?

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Historique

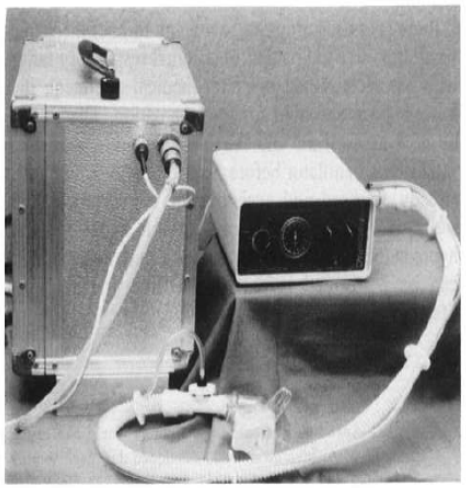
« Pont » vers la transplantation

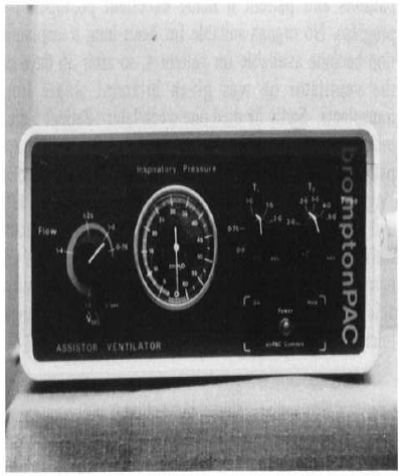
Case	Age yrs	Sex	When put on HLT waiting list		Before ventilation			On ventilator			Duration of NIPPV	Outcome
			FEV_1 *l*	FVC *l*	Inspired oxygen	Pao_2 kPa	$Paco_2$ kPa	Added oxygen *l*·min^{-1}	Pao_2 kPa	$Paco_2$ kPa	Days	
1	36	F	600	1210	Air	3.9	9.7	2	10.5	7.7	10	HLT. Extubated at 48 h. Discharged from ICU at 6 days postop.
2	21	M	800	2500	28%	3.9	11.6	2	6.9	11.3	3.5	HLT. Extubated at 36 h. Discharged from ICU at 6 days postop.
3	17	M	520	740	60%	4.3	10.9	2	8.2	8.2	3	HLT. Extubated at 6 h. Discharged from ICU at 13 days postop.
4	29	M	650	900	35%	6.5	8.4	1	10.9	8.7	36	No HLT organs available. Bilateral single lung transplant. Died one week after surgery of bronchial anastomotic problems and sepsis.
5	19	F	560	960	30%	5.3	9.9	2	9.0	8.8	15	No organs available. Died of pulmonary sepsis.
6	27	F	350	970	30%	8.99	14.87	2	13.36	11.95	17	HLT extubated at 18 h. Discharged from ICU 3 days postop.

Hodson et al, Eur Respir J, 1991

Historique

« Pont » vers la transplantation





Hodson et al, Eur Respir J, 1991

Historique

« Pont » vers la transplantation

Case	Age yrs	Sex	When put on HLT waiting list: FEV_1 *l*	When put on HLT waiting list: FVC *l*	Before ventilation: Inspired oxygen	Before ventilation: Pao_2 kPa	Before ventilation: $Paco_2$ kPa	On ventilator: Added oxygen *l*·min^{-1}	On ventilator: Pao_2 kPa	On ventilator: $Paco_2$ kPa	D	Outcome
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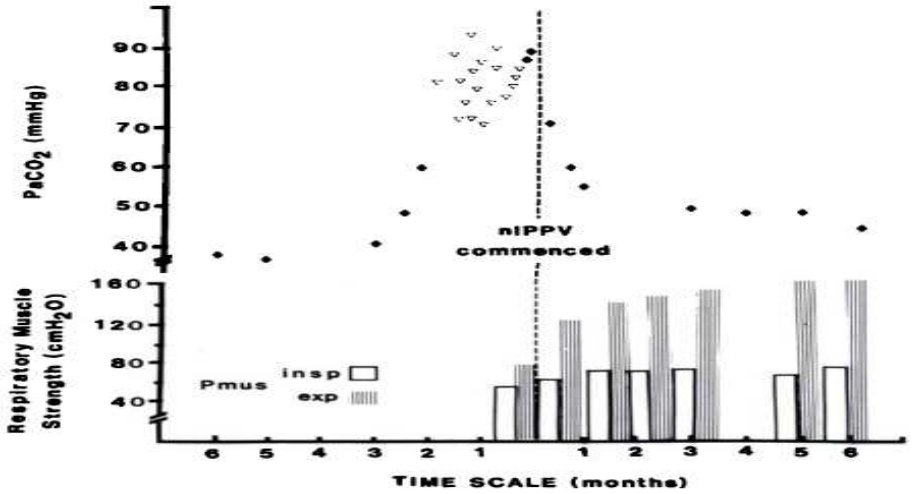
											17	

Hodson et al, Eur Respir J, 1991

Suite

Table 1—*Clinical Details of Four Patients with CF and Hypercapnic Respiratory Failure*

Patient No./Sex/Age, yr	FEV_1, L	FVC, L	FEV_1, %Pred	FVC, %Pred	NIH[12] Score	Body Mass Index, kg/sq m
1/M/25	0.55	1.2	14	25	46	18.8
2/M/30	0.55	1.45	16	30	44	18.1
3/M/29	0.5	1.8	11	36	35	17.3
4/F/25	0.55	0.69	14	16	53	17.5



Piper et al, Chest, 1992

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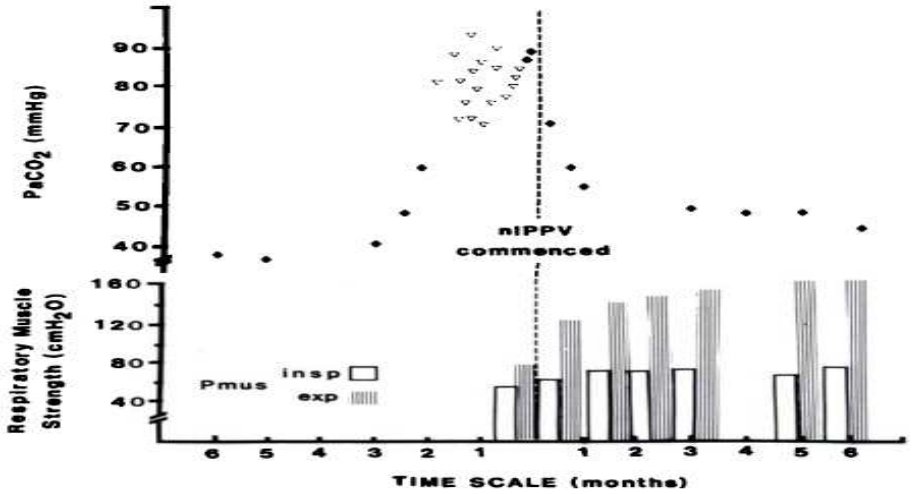


Piper et al, Chest, 1992

Confirmation

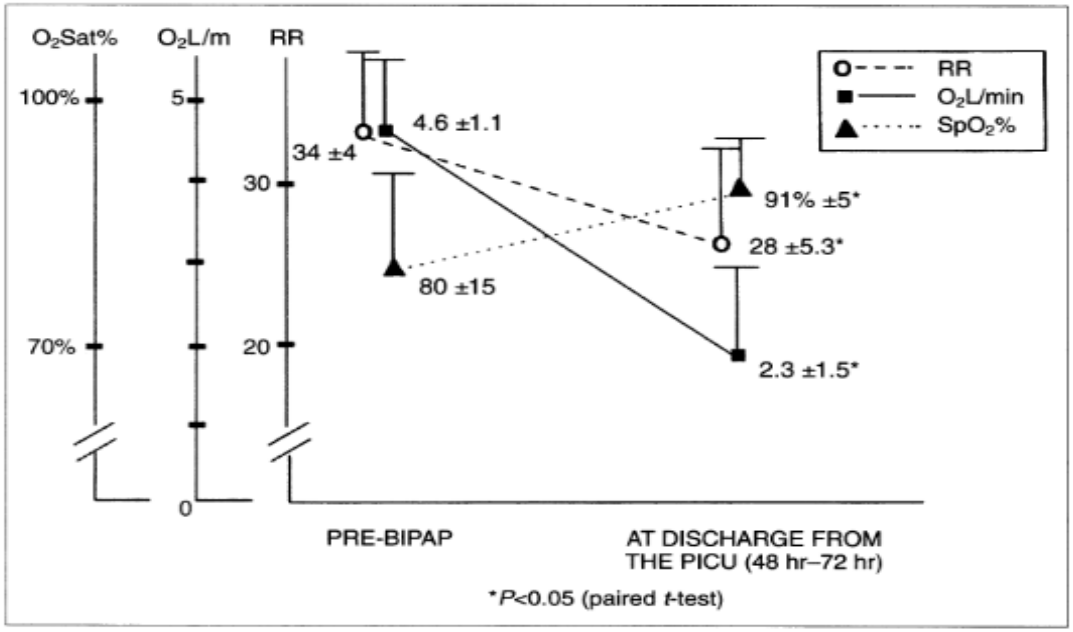
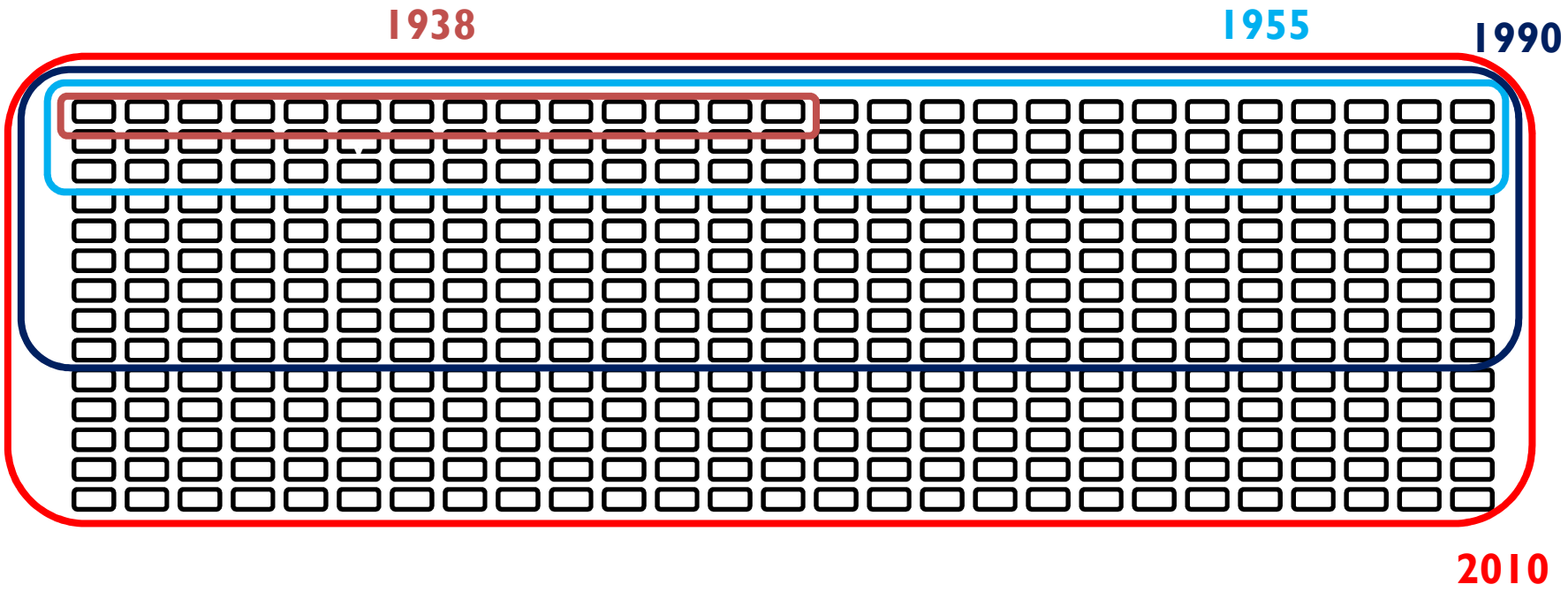


Figure 1. Pre-BIPAP and 48–72 hours post-BIPAP respiratory physiologic variables. Note the significant improvement in respiratory rate (RR), oxygen flow requirement (O_2 L/min), and oxygen saturation of hemoglobin (SpO_2) at the time of discharge from the PICU.

Caronia et al, Clin Pediatr, 1998

Indications et Espérance de Vie



1 mois

1 an

Adapté de Davis, Am J Resp Crit Care Med, 2006

Indications actuelles

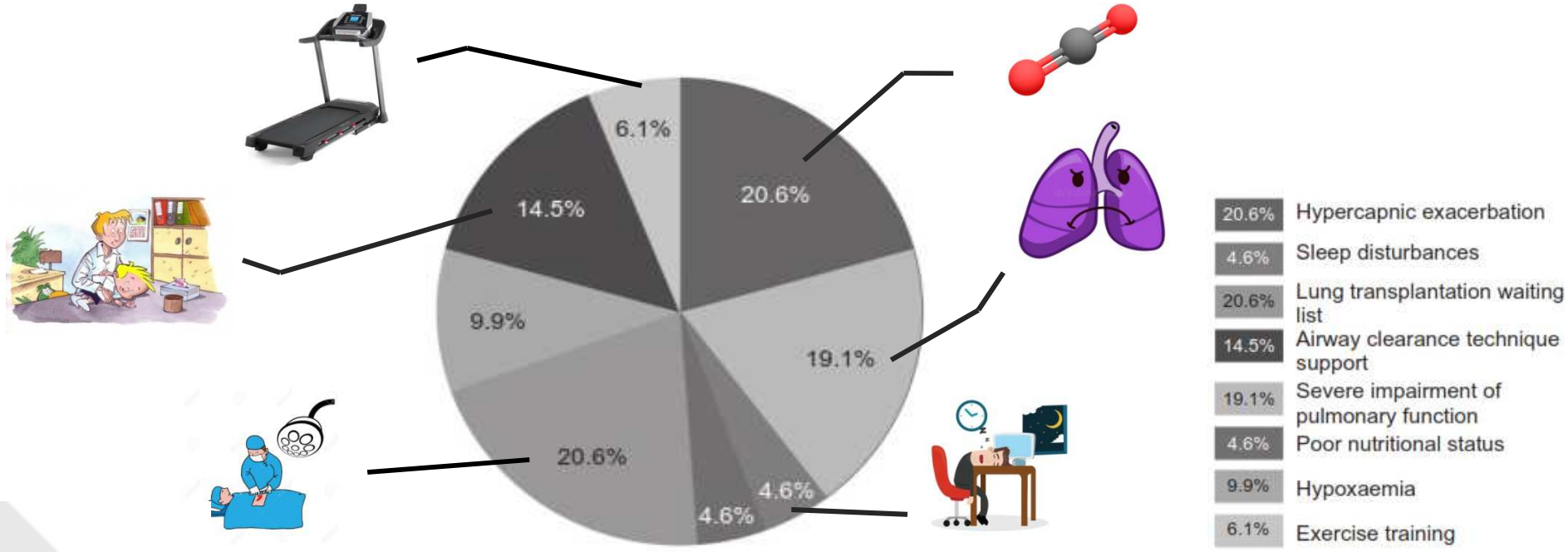
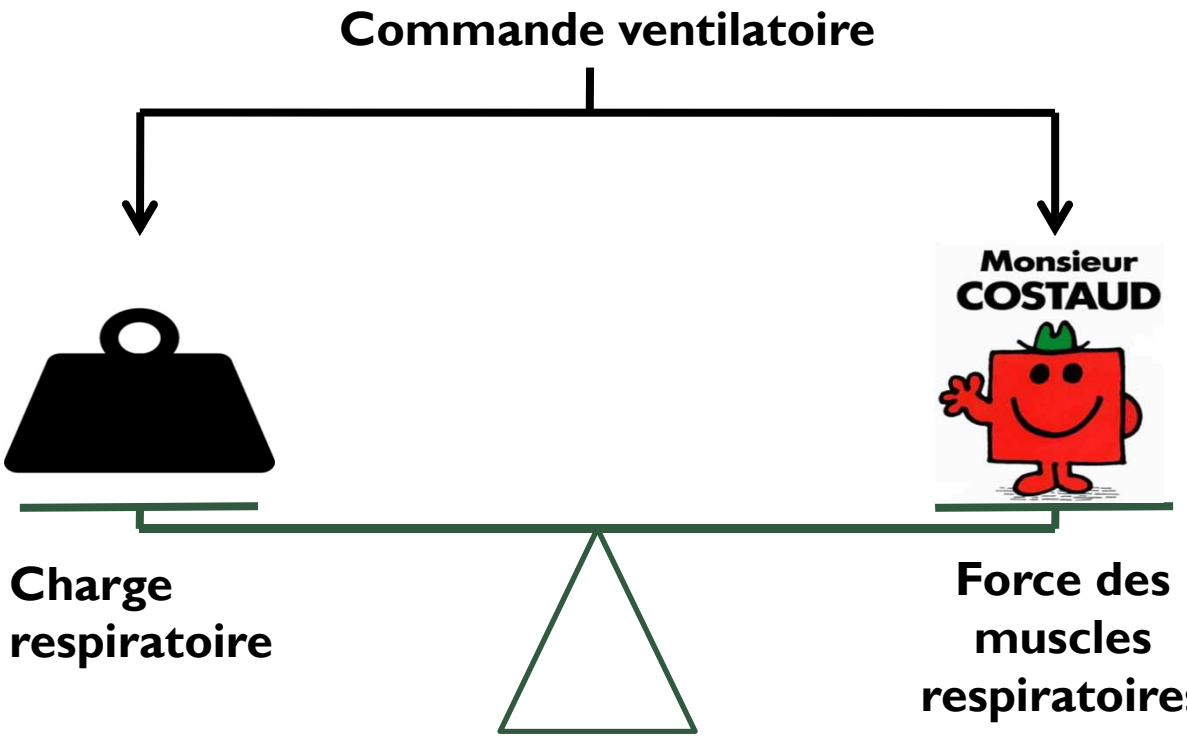


FIGURE 1. The main criteria for noninvasive ventilation initiation according to physiotherapists.

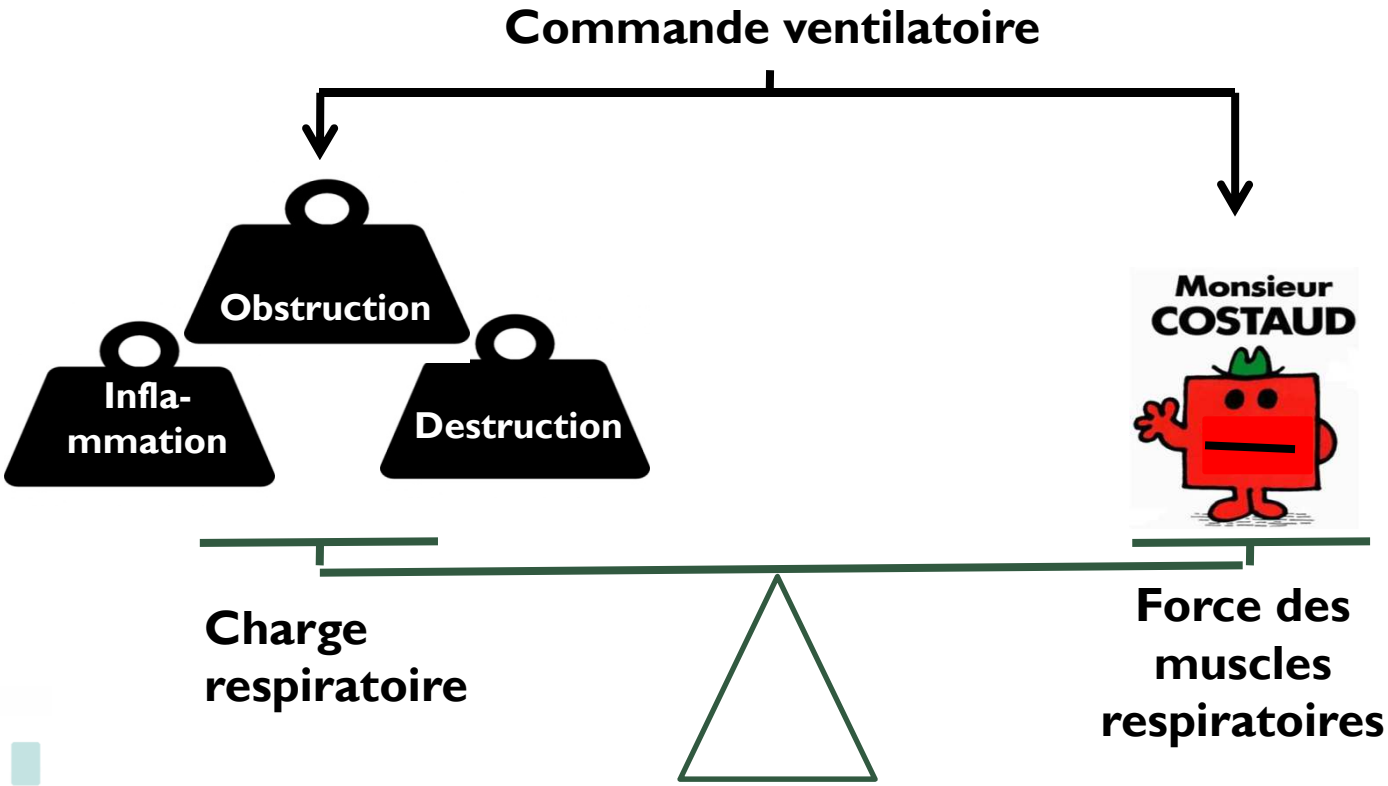
Zuffo et al, Eur Respir J, 2015

L'équilibre respiratoire



D'après Fauroux, Minerva Anestesiol, 2011

Cas de la mucoviscidose



D'après Fauroux, Minerva Anestesiol, 2011

Adaptation respiratoire

- Compensation :

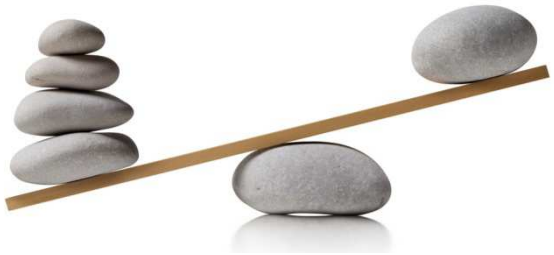
- ↗ FR
- Maintien VE
- Risque d'hypoventilation alvéolaire

  ↘ PaO_2 / ↗ $PaCO_2$

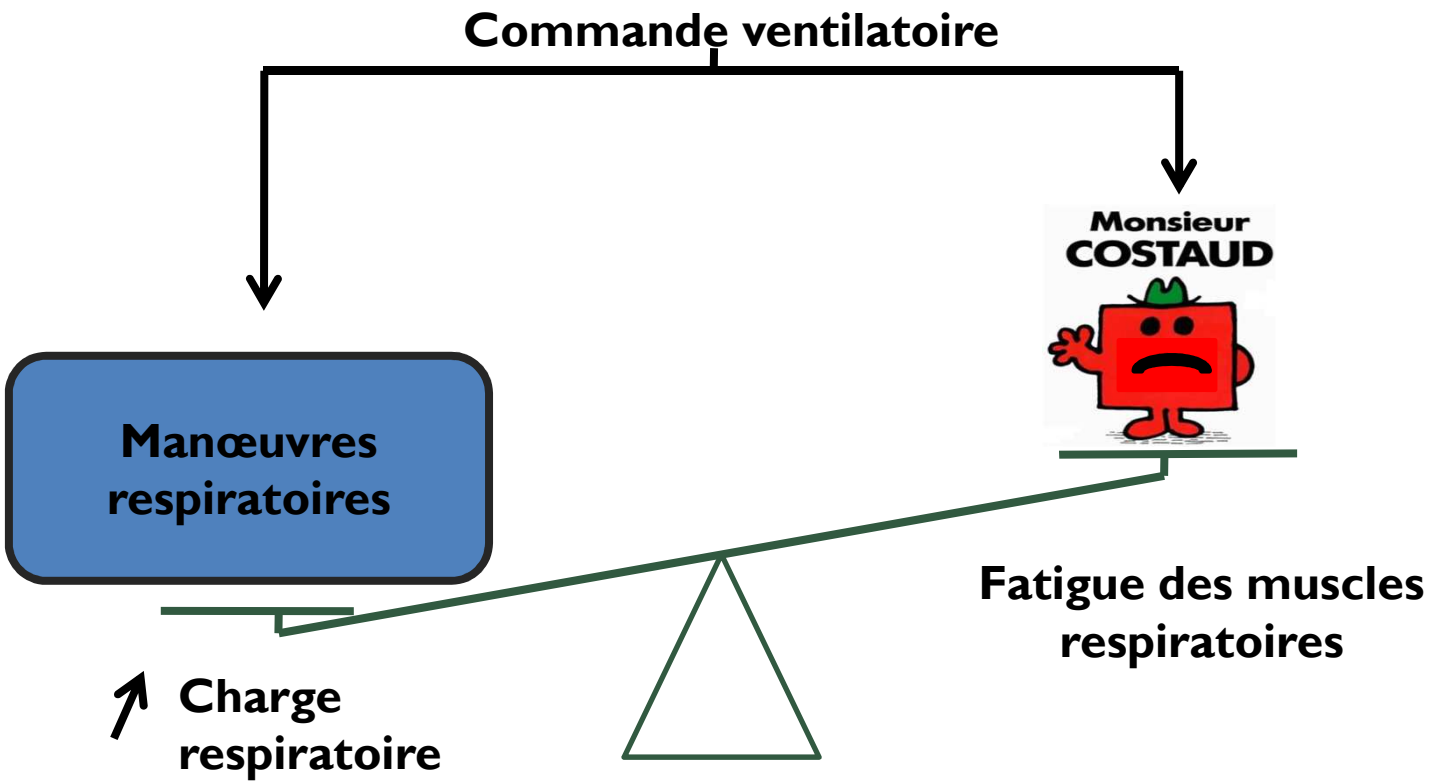
Hart et al, Am J Respir Crit Care Med, 2002

Utilisation de la VNI ?

Situations de déséquilibre dans lesquelles la compensation physiologique n'est plus suffisante …

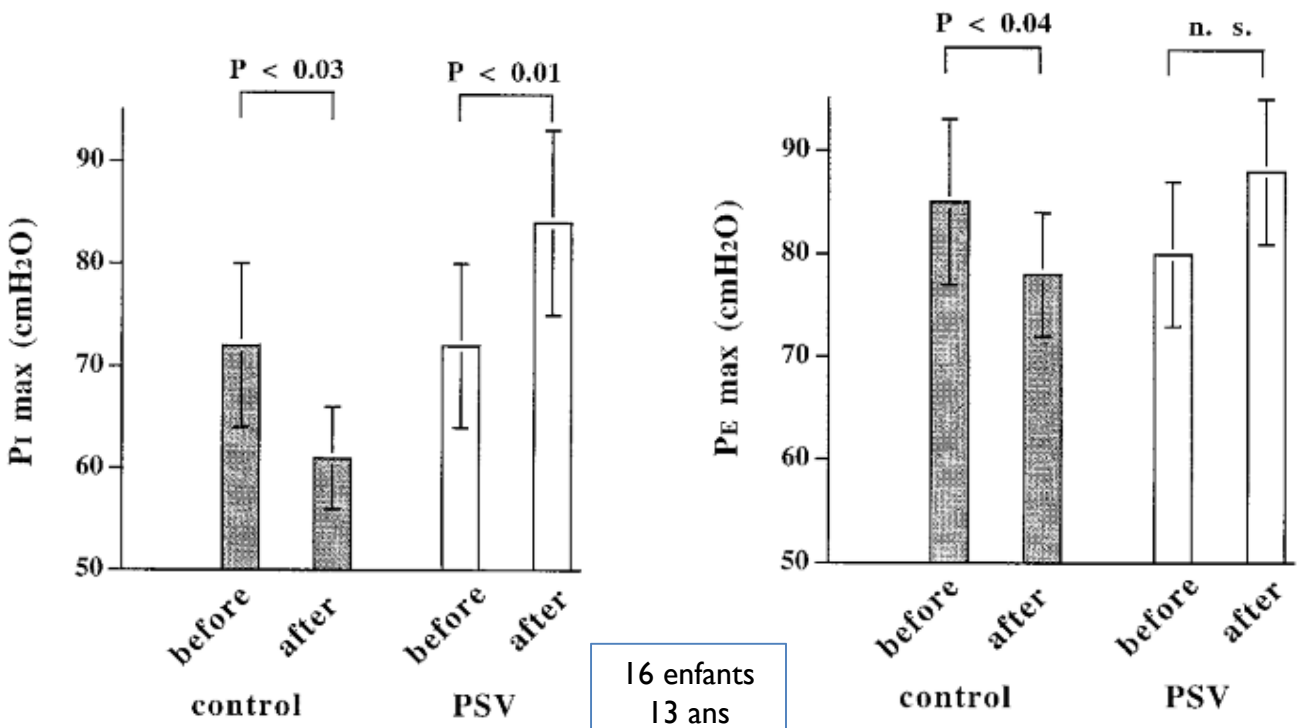


1er exemple (rapide) : Kinésithérapie respiratoire ?



D'après Fauroux, Minerva Anestesiol, 2011

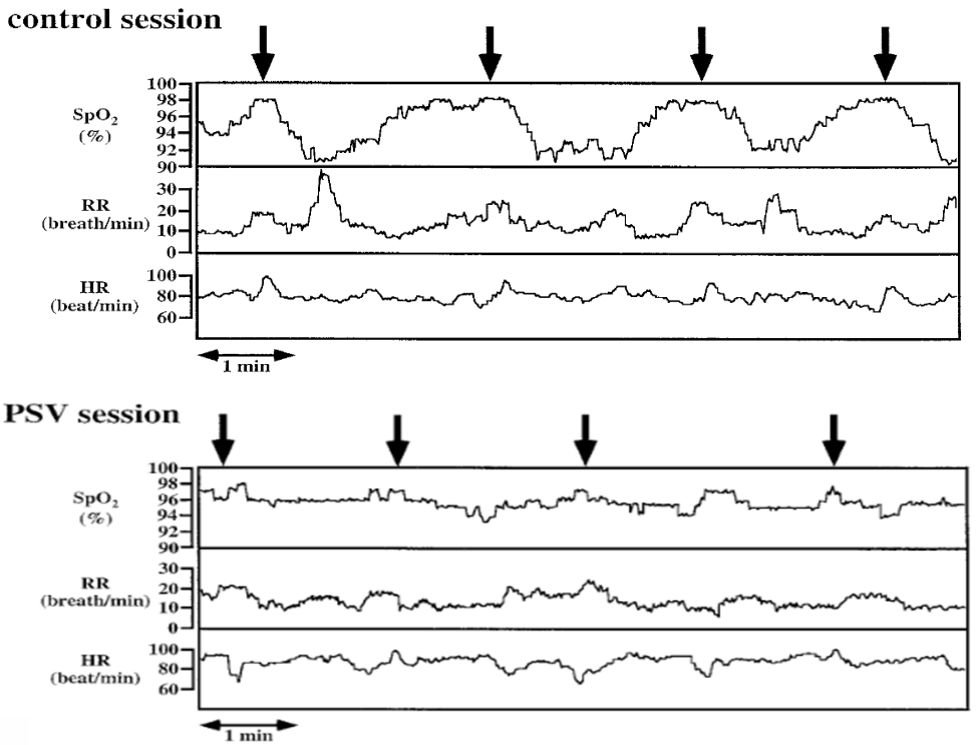
Fatigue des muscles respiratoires

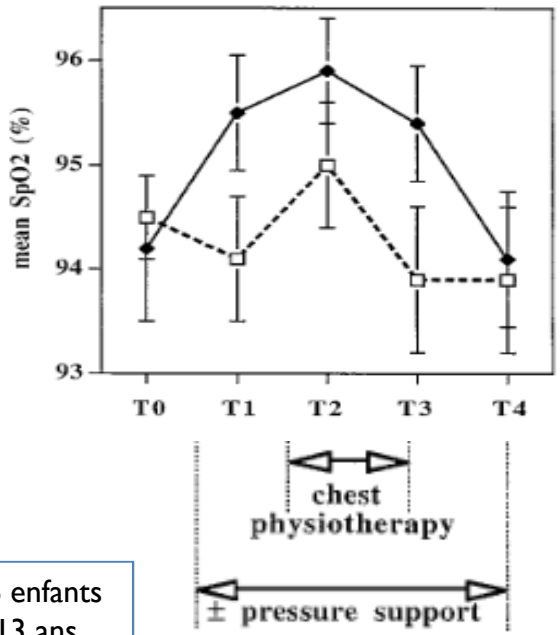


16 enfants
13 ans
50% FEV1

Fauroux et al, Pediatrics, 1999

Intérêt immédiat chez l'enfant ?





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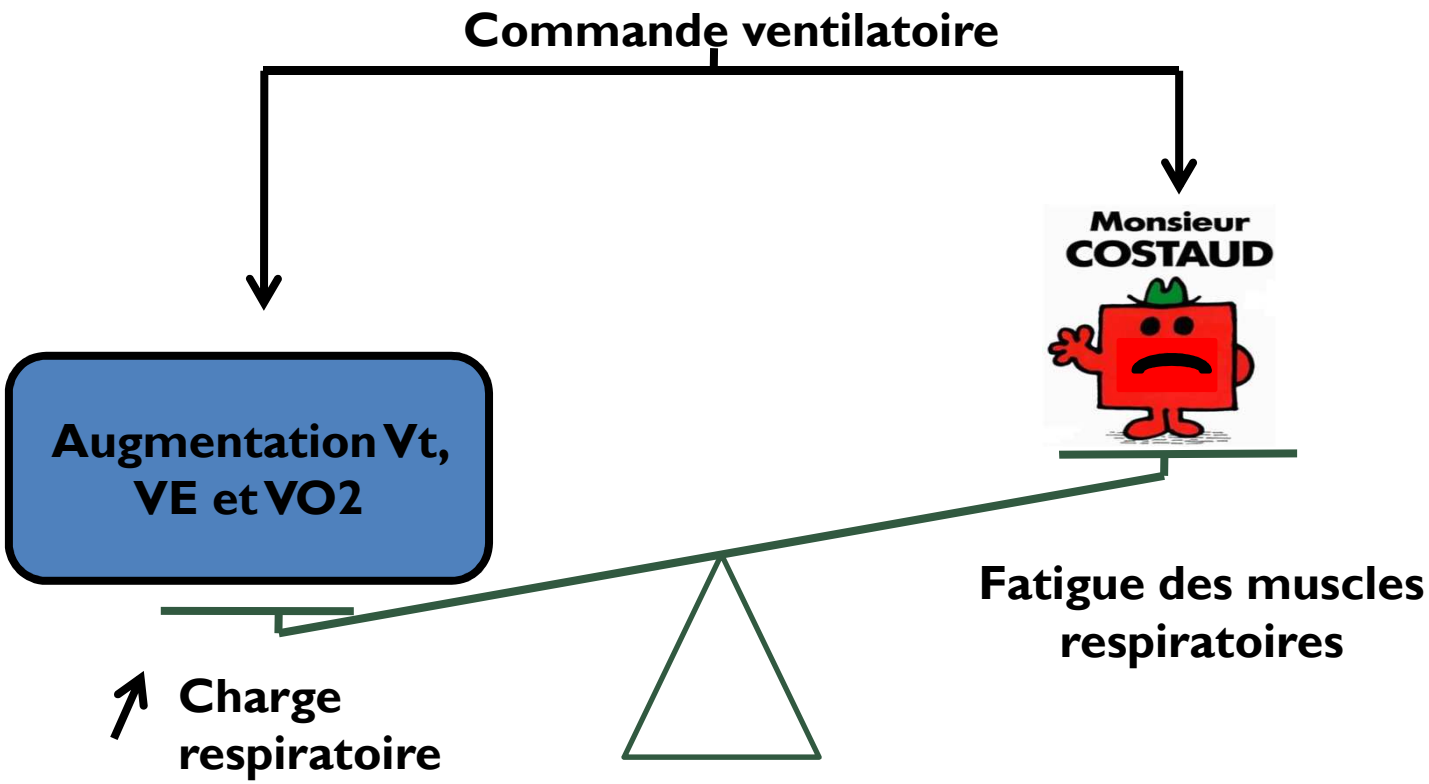
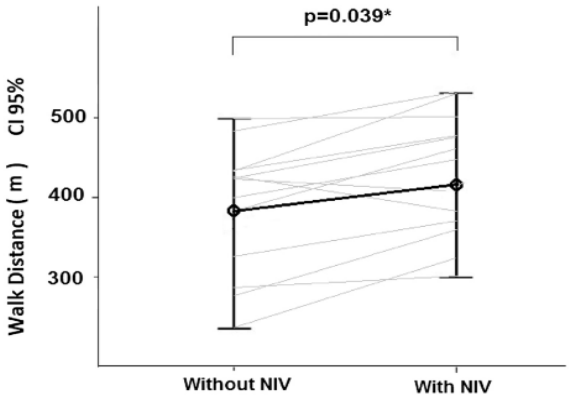
Fauroux et al, Pediatrics, 1999

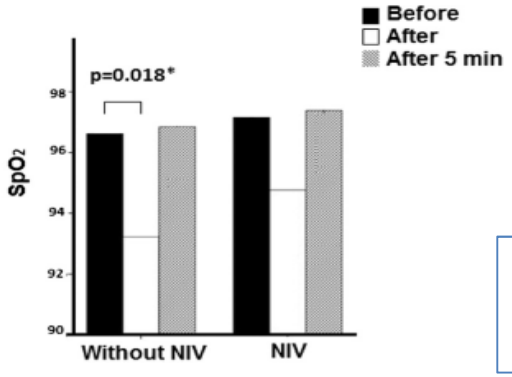
2ème exemple (rapide aussi) : Activité physique ?



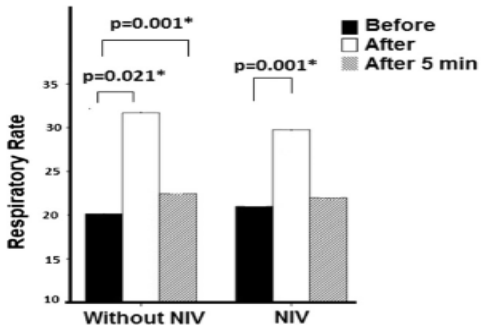
D'après Fauroux, Minerva Anestesiol, 2011

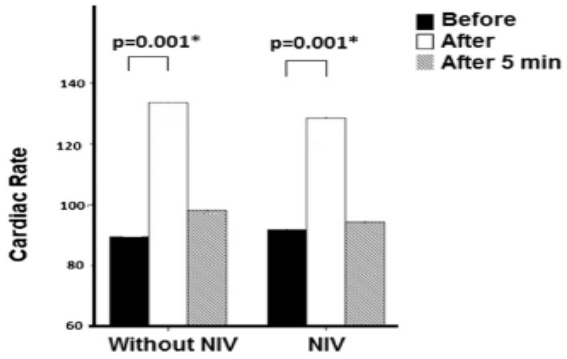
VNI et effort





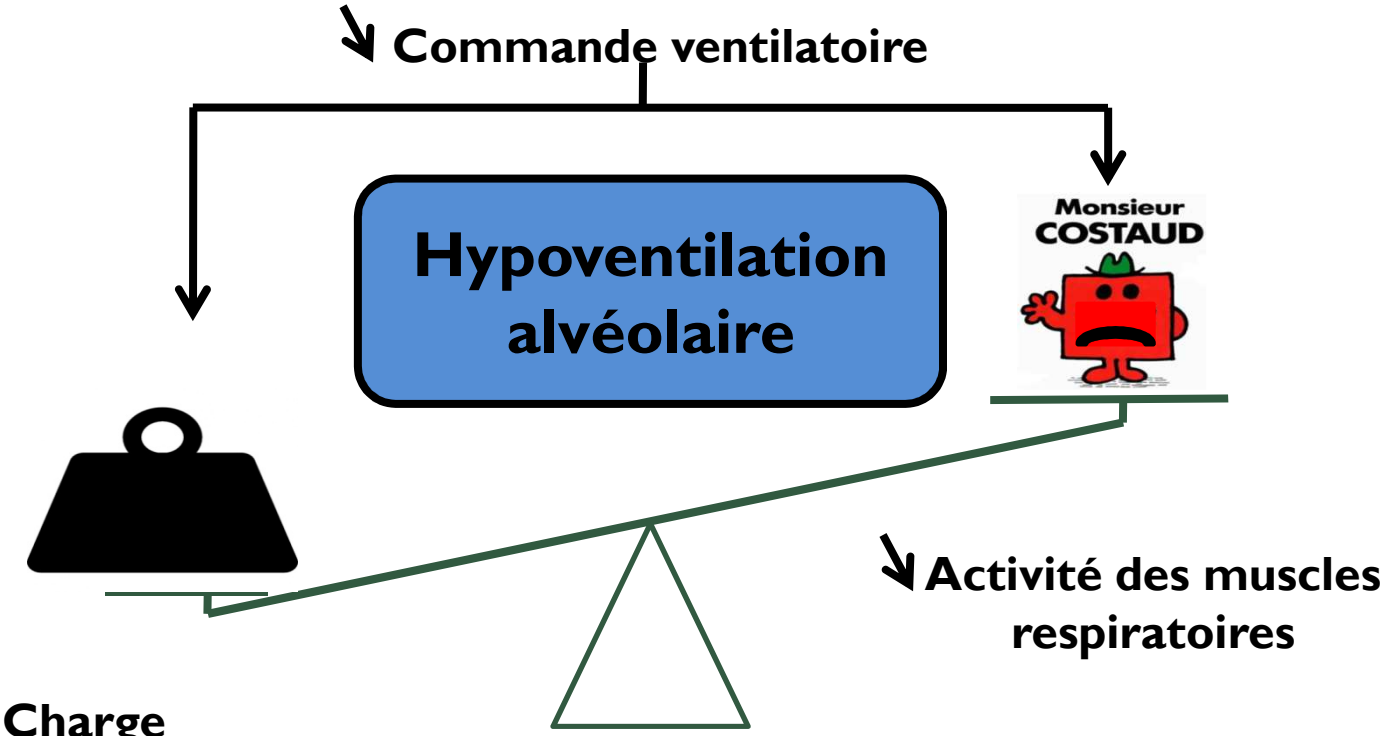
13 enfants
7-16 ans
FEV1 35-85%





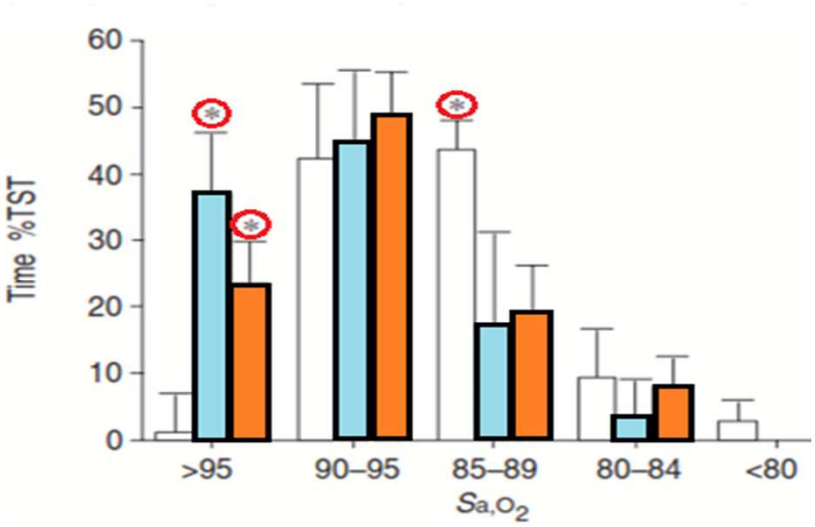
Lima et al, Respir Med, 2014

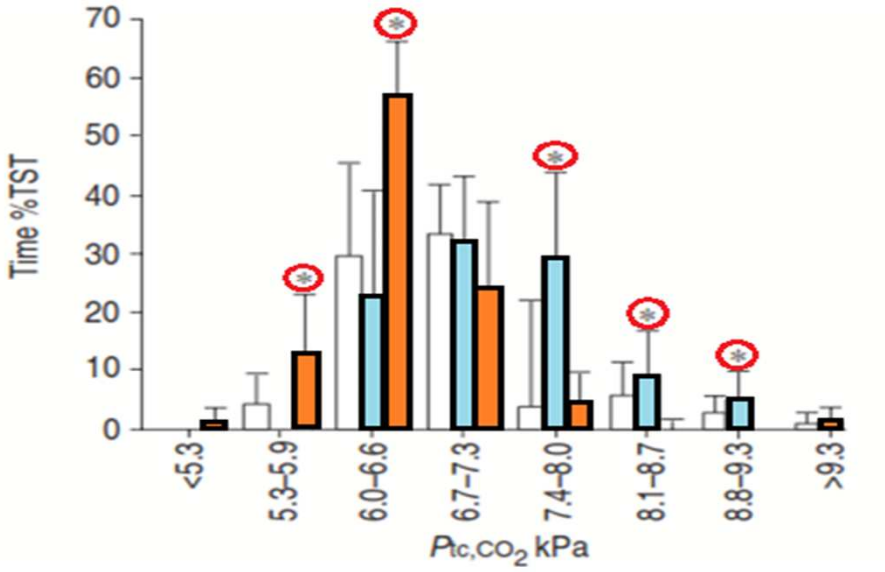
3ème exemple : Le cas du sommeil



D'après Fauroux, Minerva Anestesiol, 2011

Hypoxémie et hypercapnie



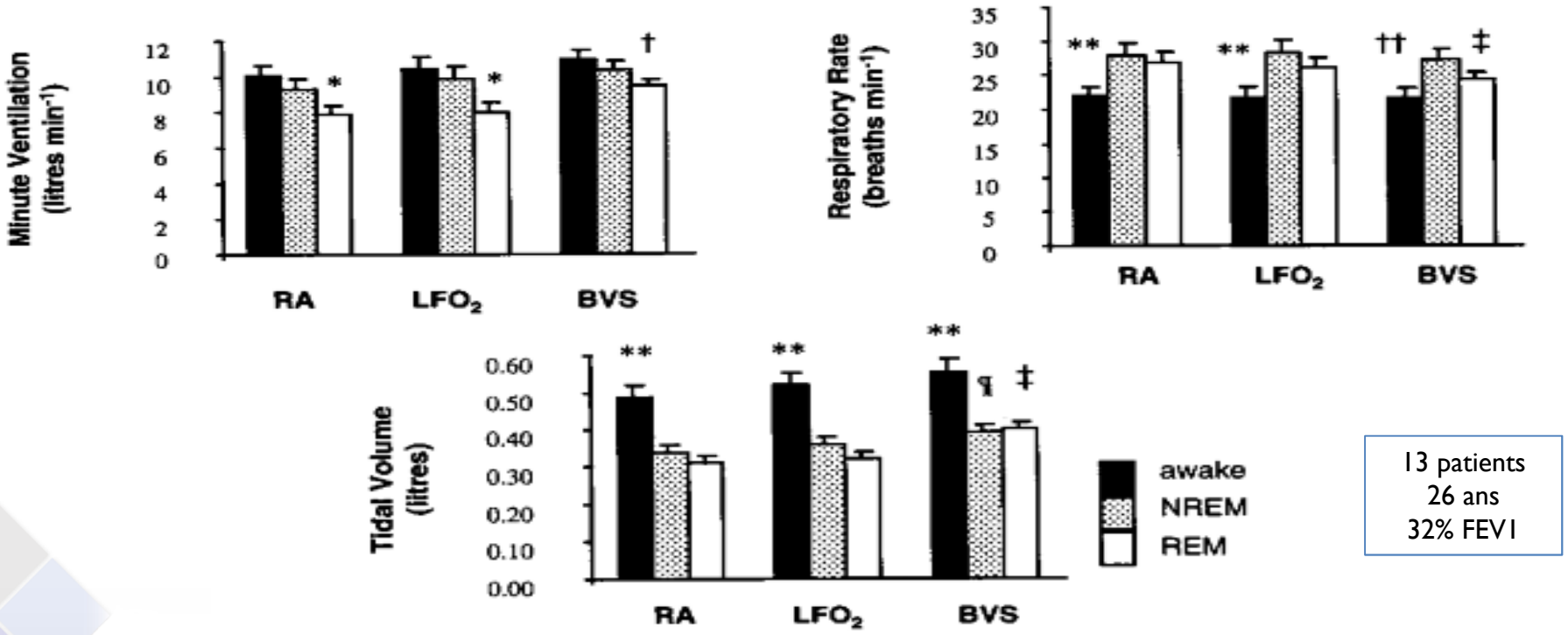


VNI
O2
Control

6 patients
22 ans
30% FEV1

D'après Gozal, Eur Respir, 1997

« Changement de rythme »



13 patients
26 ans
32% FEV1

Milross et al, Am J Respir Crit Care Med, 2001

Un sommeil plus confortable

Table 2 Quality of life

	Baseline	Air	Oxygen	NIV
CF QoL (0 = worst, 100 = best)				
Physical functioning	69 (20)	68 (22)	64 (21)	67 (19)
Chest symptoms	65 (13)	64 (20)	68 (20)	71 (17)†
Treatment issues	58 (15)	67 (20)	63 (35)	65 (21)
Emotional responses	83 (11)	85 (11)	83 (13)	79 (18)
CSQ (0 = worst, 100 = best)	42 (16)	56 (22)	50 (11)	58 (13)
ESS (0 = best, 24 = worst)	9 (5)	7 (5)	7 (6)	7 (5)
Global PSQI (0 = best, 21 = worst)	7 (3)	7 (3)	6 (2)	6 (3)
MRC (0 = best, 4 = worst)	2.4 (0.9)	2.6 (1.2)	2.5 (0.8)	2.6 (1.0)
TDI (−9 = worst, +9 = best)	5.8 (2.7)*	−1.9 (2.5)	−0.4 (1.5)	1.0 (1.8)†

8 patients
37 ans
35% FEV1
6 semaines

Young et al, Thorax, 2008

Un sommeil plus confortable

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Chest symptoms 65 (13) 64 (20) 68 (20) 71 (17)†

Moins de symptômes
Moins de dyspnée

Young et al, Thorax, 2008

Capacité à l'effort

Table 5 Arterial blood gases, pulmonary function tests and Modified Shuttle Walk Test

	Baseline	Air	Oxygen	NIV
pH	7.38 (0.02)	7.38 (0.03)	7.39 (0.02)	7.39 (0.03)
$Paco_2$ (mm Hg)	52 (4)	52 (7)	51 (7)	50 (5)
Pao_2 (mm Hg)	63 (5)	64 (7)	66 (12)	62 (6)
Hco_3 (mmol/l)	30 (2)	30 (3)	30 (4)	30 (2)
Sao_2 (%)	93 (3)	93 (3)	94 (4)	92 (4)
FEV_1 (% pred)	35 (8)	32 (10)	32 (9)	33 (9)
FVC (% pred)	60 (11)	54 (13)	54 (15)	58 (15)
RV/TLC (%)	59 (8)	64 (9)	64 (11)	63 (10)
Shuttle Walk Test (m)	434 (198)	381 (132)	403 (114)	459 (144)*

8 patients
37 ans
35% FEV1
6 semaines

Young et al, Thorax, 2008
Bradley et al, Chest, 2000

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Amélioration fonctionnelle
MCID = 40m

Young et al, Thorax, 2008
Bradley et al, Chest, 2000

Physiologie du sommeil

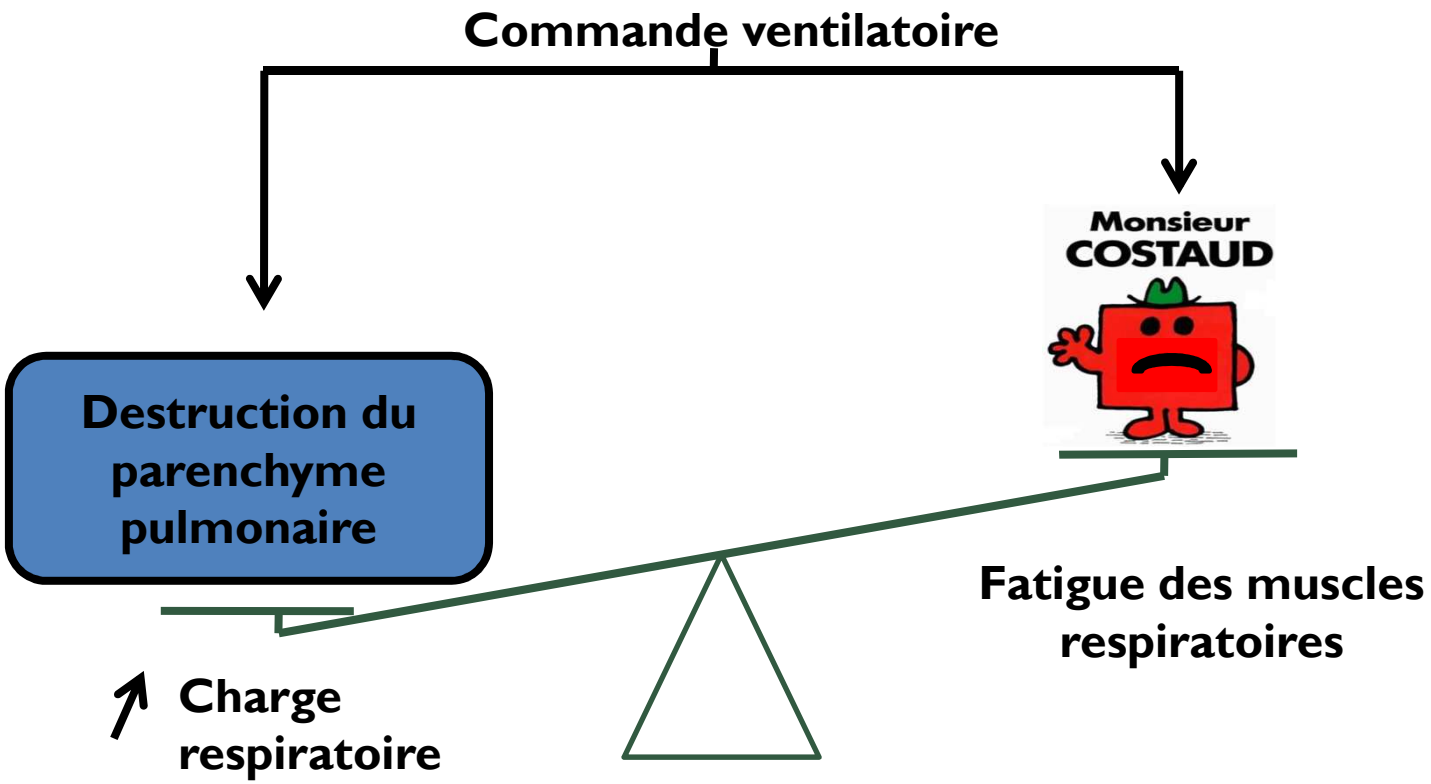
Table 4 Sleep physiology

	Baseline	Air	Oxygen	p Value*	NIV	p Value†
Mean Spo_2 for TST (%)	90 (4)	89 (5)	93 (2)	0.03	92 (3)	0.13
Minimum Spo_2 for TST (%)	76 (9)	75 (15)	82 (6)	0.24	79 (9)	0.42
Min av Spo_2 for TST (%)	89 (4)	88 (6)	92 (1)	0.03	91 (2)	0.12
TST with Spo_2 <90% (%)	38 (43)	48 (50)	10 (9)	0.03	23 (34)	0.13
Mean $Ptcco_2$ TST (mm Hg)	58 (3)	62 (13)	62 (10)	0.92	53 (6)	0.02
Max $Ptcco_2$ TST (mm Hg)	68 (12)	74 (19)	72 (11)	0.62	58 (7)	0.005‡
$\Delta Ptcco_2$ (mm Hg)	4.5 (1.6)	4.9 (2.6)	5.5 (3.6)	0.45	2.7 (1.5)	0.02‡
$\Delta Paco_2$ (mm Hg)	1.3 (3.9)	2.0 (2.3)	6.0 (4.2)	0.07	−1.3 (4.1)	0.07‡
Mean RR (breaths/min)	23 (6)	23 (5)	22 (3)	0.83	23 (5)	0.74
Mean HR (beats/min)	78 (15)	73 (11)	70 (18)	0.37	64 (15)	0.05

8 patients
37 ans
35% FEV1
6 semaines

Young et al, Thorax, 2008

4ème exemple: Détresse respiratoire chronique



D'après Fauroux, Minerva Anestesiol, 2011

Détresse respiratoire chronique

Practice of Noninvasive Ventilation for Cystic Fibrosis: A Nationwide Survey in France

RESPIRATORY CARE • NOVEMBER 2008 VOL 53 NO 11

Table 2. Reported Indications for NIV for Patients With Cystic Fibrosis in 36 Participating Centers

Indication*	Pediatric Centers (n = 15)	Pediatric and Adult Centers (n = 8)	Adult Centers (n = 13)	P
Lung function				
Diurnal hypercapnia (%)	93	88	92	.82
if P_{aCO_2} more than this value†	63 (61–64 mm Hg)	50 (45–58 mm Hg)	48 (45–50 mm Hg)	.78
According to a given FEV_1 % predicted value	40	25	15	.72
if FEV_1 % predicted less than this value†	35 (32–35%)	25 (22–28%)	30 (25–35%)	.36

36 centres
4.416 patients

Fauroux et al, Respir Care, 2008

Une détérioration « plus lente »

	Pre-NIPPV	1–2 months	Pre-NIPPV	3 months	Pre-NIPPV	6 months
Number	10	10	5	5	3	3
FEV_1 (%)	15·4 ± 1·3	20·9 ± 3·3*	16·8 ± 2·2	20·6 ± 3·4	14·8 ± 3·3	17·5 ± 4·6
FVC (%)	26·5 ± 4·1	31·9 ± 4·4*	29·2 ± 5·9	35 ± 5·5*	23·4 ± 5·7	28·8 ± 8
BMI (kg m^{-2})	16·8 ± 0·5	17·5 ± 0·5	17·5 ± 1	17·1 ± 1	17·7 ± 1·2	17·2 ± 1·5
$PaCO_2$ (kPa)	8·5 ± 0·5	8·5 ± 0·6	8·5 ± 1·1	7·5 ± 0·7*	8·7 ± 1·9	6·4 ± 1
PaO_2 (kPa)	7·3 ± 0·7	6·9 ± 0·5	7·7 ± 1	7·5 ± 0·7	8·7 ± 1	10 ± 1·3
$[HCO_3^-]$ (mmol l^{-1})	40·4 ± 1·7	40·7 ± 2·3	39·7 ± 2·2	34·1 ± 1·5*	37·9 ± 2·8	31·7 ± 3·2

FEV_1 (%), percentage predicted forced expired volume in 1 s; FVC (%), percentage predicted forced vital capacity; BMI, body mass index; $PaCO_2$, arterial concentration of carbon dioxide; PaO_2, arterial concentration of oxygen; $[HCO_3^-]$, arterial concentration of bicarbonate.
*$P<0·05$ compared with patients pre-NIPPV.

12 patients
26 ans
15% FEV1

Hill et al, Respir Med, 1998

Une détérioration « plus lente »

	Pre-NIPPV	1–2 months	Pre-NIPPV	3 months	Pre-NIPPV	6 months
Number	10	10	5	5	3	3
FEV_1 (%)	15·4 ± 1·3	20·9 ± 3·3*	16·8 ± 2·2	20·6 ± 3·4	14·8 ± 3·3	17·5 ± 4·6
FVC (%)	26·5 ± 4·1	31·9 ± 4·4*	29·2 ± 5·9	35 ± 5·5*	23·4 ± 5·7	28·8 ± 8
$PaCO_2$ (kPa)	8·5 ± 0·5	8·5 ± 0·6	8·5 ± 1·1	7·5 ± 0·7*	8·7 ± 1·9	6·4 ± 1
PaO_2 (kPa)	7·3 ± 0·7	6·9 ± 0·5	7·7 ± 1	7·5 ± 0·7	8·7 ± 1	10 ± 1·3
$[HCO_3^-]$ (mmol l^{-1})	40·4 ± 1·7	40·7 ± 2·3	39·7 ± 2·2	34·1 ± 1·5*	37·9 ± 2·8	31·7 ± 3·2

FEV_1 (%), percentage predicted forced expired volume in 1 s; FVC (%), percentage predicted forced vital capacity; BMI, body mass index; $PaCO_2$, arterial concentration of carbon dioxide; PaO_2, arterial concentration of oxygen; $[HCO_3^-]$, arterial concentration of bicarbonate.
*$P<0·05$ compared with patients pre-NIPPV.

12 patients
26 ans
15% FEV1

Hill et al, Respir Med, 1998

Une inversion/stabilisation de l'effet ?

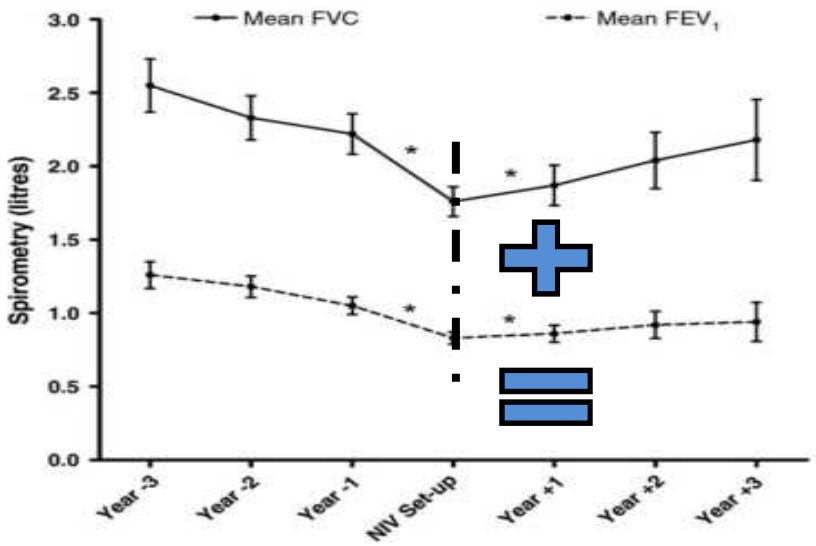
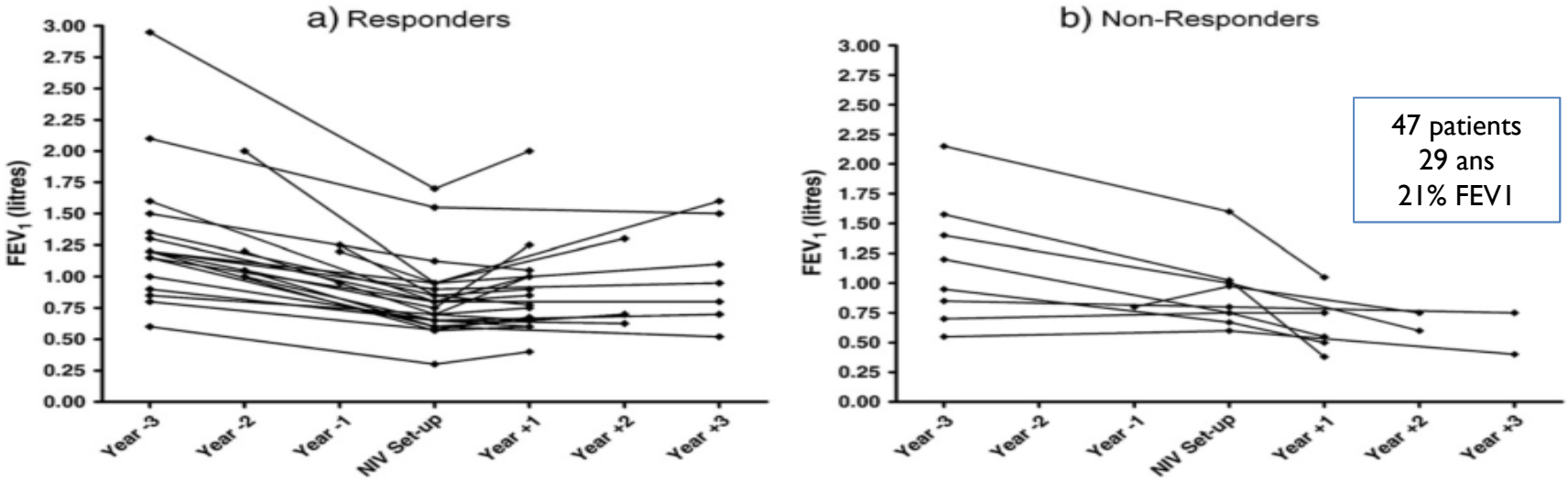


Fig. 1. Effect of long-term NIV on mean forced expiratory volume in 1 s (FEV_1) and forced vital capacity (FVC). Error bars represent standard deviation. * $p<0.01$.

47 patients
29 ans
21% FEV1

Flight et al, J Cyst Fibros, 2012

Avec ses limites

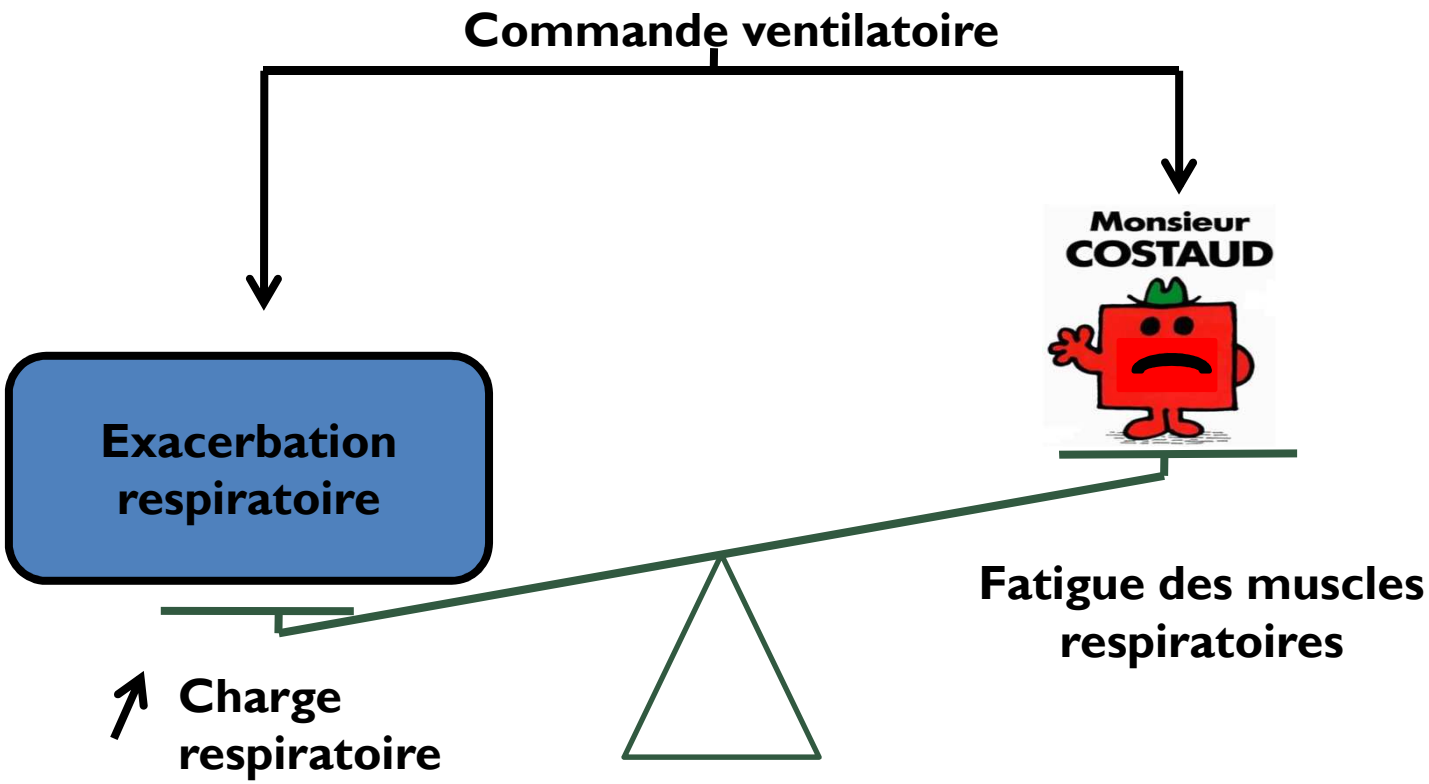


47 patients
29 ans
21% FEV1

VEMS pré-NIV : -195mL vs -61mL (p<0,01)

Flight et al, J Cyst Fibros, 2012

5ème exemple: Exacerbation (DRA)



D'après Fauroux, Minerva Anestesiol, 2011

Exacerbation

Practice of Noninvasive Ventilation for Cystic Fibrosis: A Nationwide Survey in France

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Table 2. Reported Indications for NIV for Patients With Cystic Fibrosis in 36 Participating Centers

Indication*	Pediatric Centers (n = 15)	Pediatric and Adult Centers (n = 8)	Adult Centers (n = 13)	P
Exacerbation				
During an exacerbation (%)	87	88	100	.30
After an exacerbation (%)	47	63	31	.07
According to a given P_{aCO_2} value (%)	87	63	100	.18
if P_{aCO_2} more than this value†	45 (45–55 mm Hg)	45 (45–50 mm Hg)	45 (45–50 mm Hg)	.90

36 centres
4.416 patients

Fauroux et al, Respir Care, 2008

VNI en exacerbation

Référence	Utilisation de la VNI	Taux de réussite	Taux d'échec et devenir
Ellafi et al, Am J Resp Crit Care Med, 2005	61% en ICU 33% en pneumologie	**83%** ICU **100%** pneumologie	4/23 (17%) ICU, †
Sood et al, Am J Resp Crit Care Med, 2001	13% (18 sur 136 admissions en ICU)	**78%**	4/18 (22%) †
Texereau et al, Resp Research, 2006	57% en ICU	**67%** (durée moyenne 7J)	11/34 : 8†

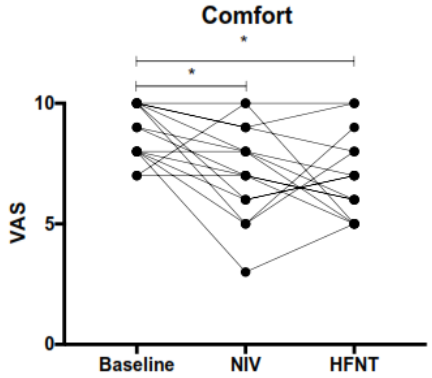
Ellafi et al, Am J Resp Crit Care Med, 2005
Sood et al, Am J Resp Crit Care Med, 2001
Texereau et al, Resp Research, 2006

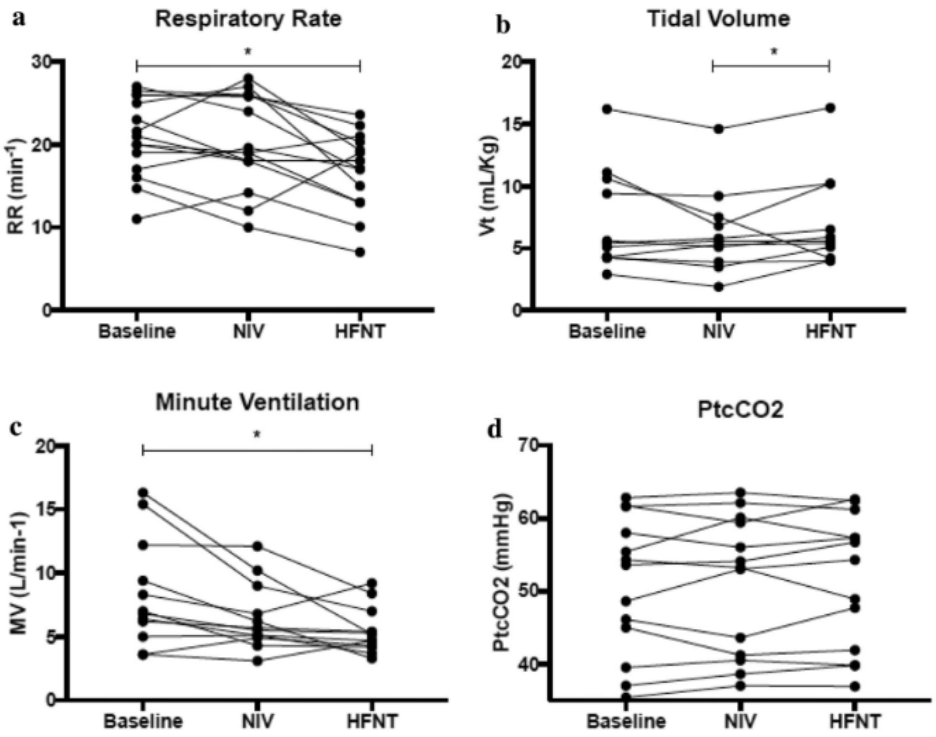
Alternative ?

RESEARCH Open Access

High-flow nasal oxygen versus noninvasive ventilation in adult patients with cystic fibrosis: a randomized crossover physiological study

15 patients
30 ans
FEV1 24%





Sklar et al, Ann Intensive Care, 2018

Limites de la VNI

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Problems observed*				
Difficulty sleeping with NIV	60	88	85	.40
Preference for oxygen therapy	13	25	15	.73
No subjective benefit	60	50	38	.44
Too constraining	47	75	77	.37
Problems with the interface	73	63	54	.64
Refusal	27	13	54	.88
Abdominal distension	17	0	54	.04
Other‡	0	0	34	.21

36 centres
4.416 patients

Fauroux et al, Respir Care, 2008

Take-Home Messages

- Complément à la KR ou à l'activité physique ?
- Correction liée à l'hypoventilation nocturne
- DRC / DRA
- Communication primordiale (*Dellon, Chest, 2012)*
- Intérêt du patient vs. « Gêne » occasionnée

Merci de votre attention

AMK
Association Mucoviscidose et Kinésithérapie

VAINCRE LA MUCOVISCIDOSE

Compléments intéressants

Cochrane Library

Cochrane Database of Systematic Reviews

Non-invasive ventilation for cystic fibrosis (Review)

Moran F, Bradley JM, Piper AJ

PHYSIOTHERAPY THEORY AND PRACTICE
https://doi.org/10.1080/09593985.2017.1400137

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Check for updates

Experience of using non-invasive ventilation as an adjunct to airway clearance techniques in adults with cystic fibrosis–A qualitative study

Maria Cecilia Rodriguez Hortal, PT, MSc[a,b,c], Anna Hedborg, PT, MSc[a,c], Gabriele Biguet, PT, MSc[d], and Malin Nygren-Bonnier, PT, PhD [a,d]

CHEST Original Research
PEDIATRICS

Physician Practices for Communicating With Patients With Cystic Fibrosis About the Use of Noninvasive and Invasive Mechanical Ventilation

Elisabeth P. Dellon, MD, MPH; Gregory S. Sawicki, MD, MPH; Mitchell D. Shores, BA; Joanne Wolfe, MD, MPH; and Laura C. Hanson, MD, MPH