2nd INTERNATIONAL PEDIATRIC NONINVASIVE VENTILATION CONFERENCE

November 3rd & 4th 2016

Is there an optimal ventilatory mode for neuromuscular children ?

Brigitte Fauroux & Alessandro Amaddeo
Pediatric noninvasive ventilation and sleep unit
Research unit INSERM U 955
Necker university Hospital, Paris, France

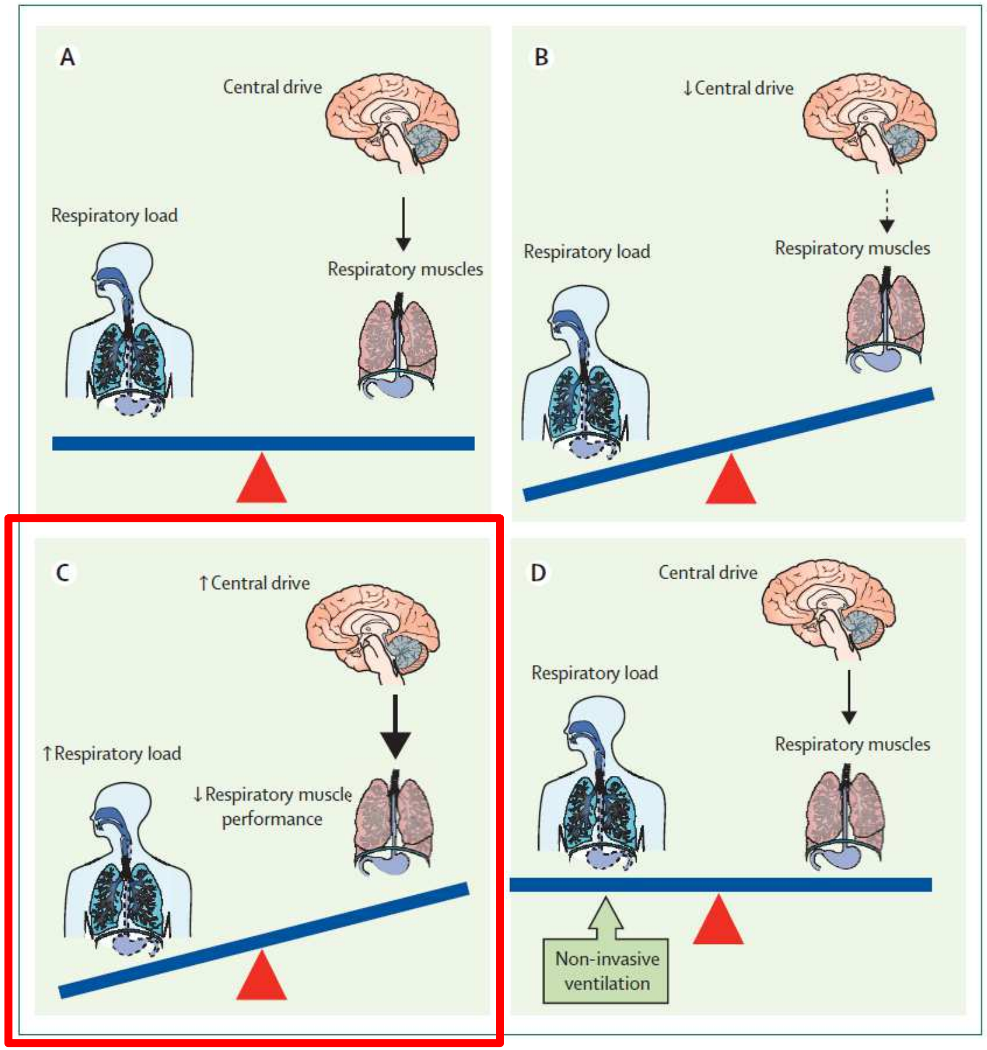
Inserm
Institut national
de la santé et de la recherche médicale

Is there an optimal ventilatory mode for neuromuscular children ?

- Introduction
- Ventilatory modes
- In practice
- Conclusion

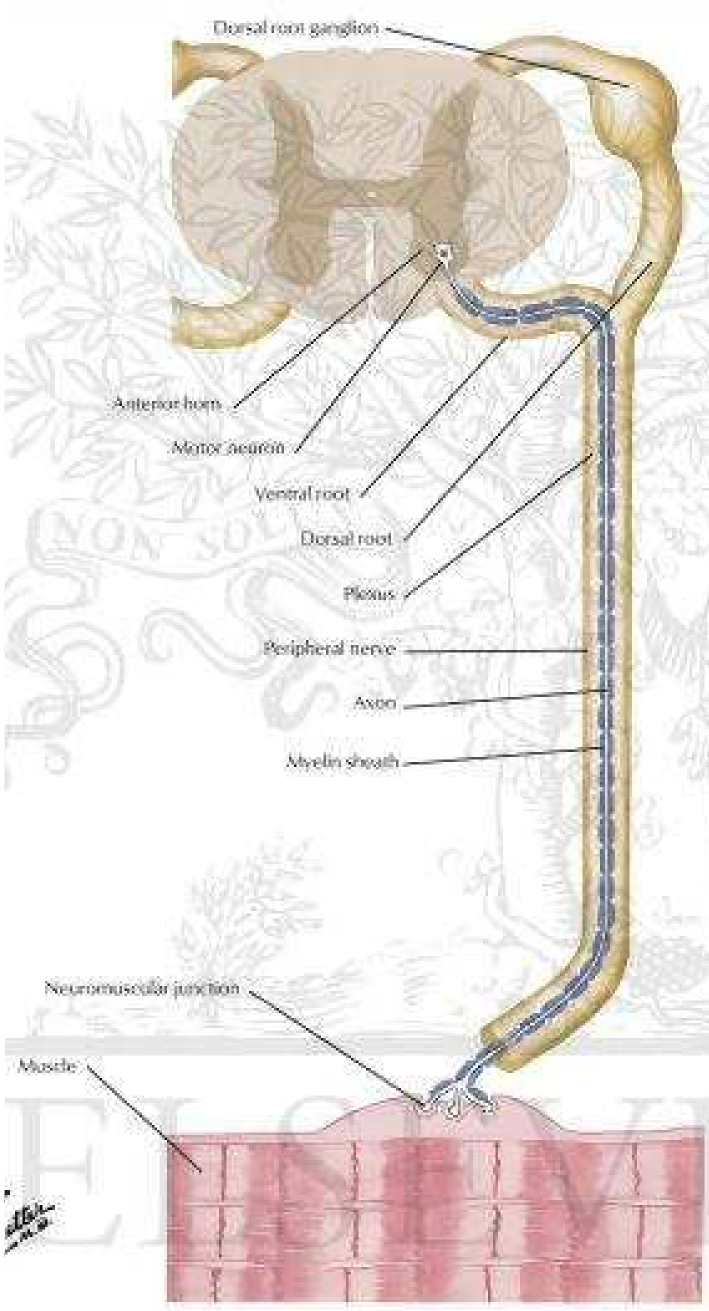
Long-term non-invasive ventilation in children

Alessandro Amaddeo, Annick Frapin, Brigitte Fauroux



www.thelancet.com/respiratory Published online July 13, 2016 http://dx.doi.org/10.1016/S2213-2600(16)30151-5

Heterogeneity of neuromuscular disorders in children



Motoneuron
- SMA

Peripheral nerve
- Metabolic
- Hereditary

Neuromuscular junction
- Myasthenia

Muscle
- DMD, myotonic D.
- Cong. myopathies

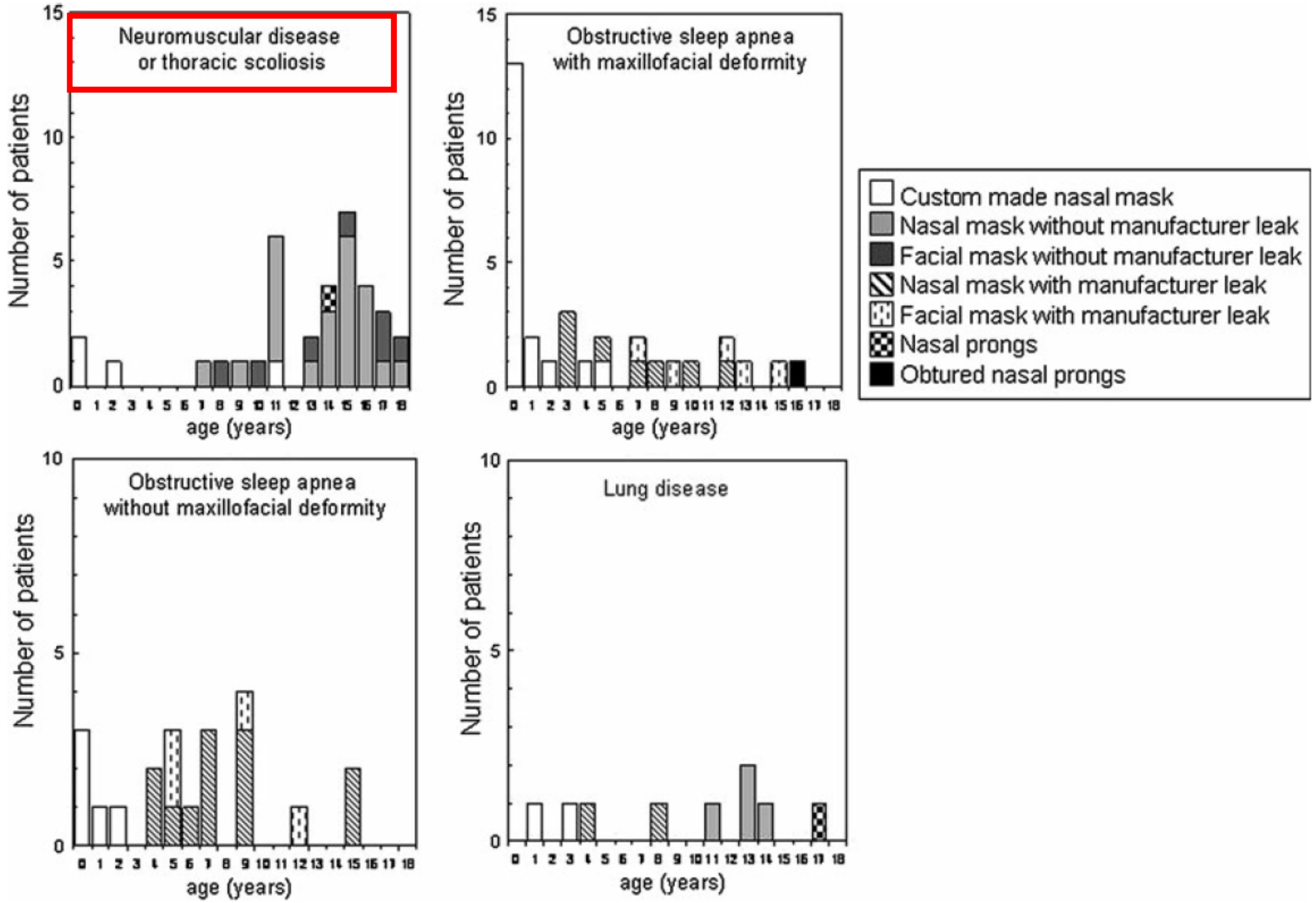
Intensive Care Med (2012) 38:655–662
DOI 10.1007/s00134-012-2516-1

PEDIATRIC ORIGINAL

Adriana Ramirez
Vincent Delord
Sonia Khirani
Karl Leroux
Sophie Cassier
Natacha Kadlub
Guillaume Aubertin
Arnaud Picard
Brigitte Fauroux

Interfaces for long-term noninvasive positive pressure ventilation in children

36%



Long-term ventilation in children: longitudinal trends and outcomes

Catherine M McDougall,[1] Robert J Adderley,[2] David F Wensley,[1,2] Michael D Seear[1,2]

Table 1 Details of long-term ventilation (LTV) patients by epoch of initiation of LTV

	Number of patients (% of total)		
	1995–1999 (n=17)	2000–2004 (n=53)	2005–2009 (n=74)
Mode of support			
Tracheostomy ventilation	6 (35)	9 (17)	13 (18)
Non-invasive CPAP	0 (0)	5 (9)	17 (23)
Non-invasive bilevel support	11 (65)	39 (74)	44 (59)
Time dependent on ventilation			
24 h/d	2 (12)	6 (11)	9 (12)
<24 h/d	15 (88)	47 (89)	65 (88)
Diagnostic category			
Neuromuscular disease	11 (64)	30 (57)	26 (35)
Spinal injury	1 (6)	3 (6)	5 (7)
Abnormal ventilatory control	1 (6)	7 (13)	11 (15)
Airway malacia	2 (12)	3 (6)	6 (8)
Craniofacial/OSA	1 (6)	6 (11)	16 (22)
Other	1 (6)	4 (7)	10 (13)
Trigger for initiation of LTV*			
Failure to wean from ventilation	5 (29)	5 (10)	18 (25)
Acute illness	6 (35)	9 (17)	16 (22)
Sleep study results	2 (12)	14 (26)	24 (33)
Symptoms of sleep-disordered breathing	3 (18)	13 (26)	7 (10)
FVC<20%	0 (0)	7 (13)	2 (3)
Other	1 (6)	4 (8)	5 (7)

McDougall *et al.* Arch Dis Childh 2013;98:660

NIV for children with NMD in Rome

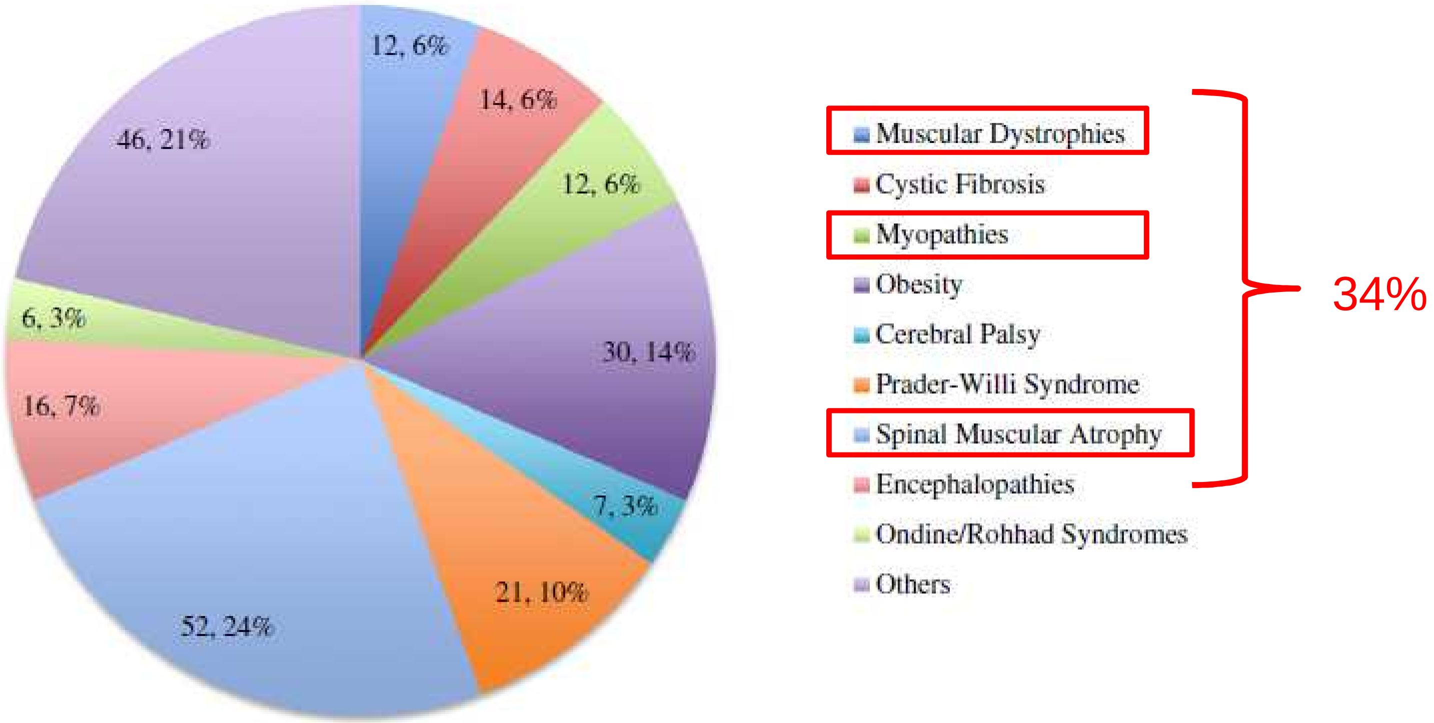


Fig. 1. Indications for non-invasive ventilation: authors' experience at Bambino Gesù Children's Research Institute (Rome, Italy).

Pavone *et al*. Early Human Development 2013;89:S25

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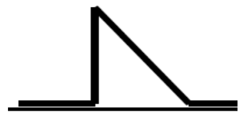
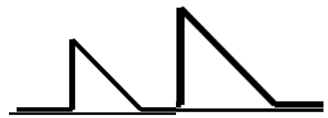
	Volume targeted ventilation	Pressure targeted ventilation	Target volume with variable pressure support
Tidal volume	stable	variable	« guaranteed »
Pressure	variable	stable	variable (within targets)
Flow			
Avantages	known tidal volume	known IPAP leak compensation	control of IPAP leak compensation « guantanteed » tidal volume
Limitations	no leak compensation no control of IPAP	variable tidal volume	possibility to guarantee tidal volume……

Paediatric Respiratory Reviews 18 (2016) 73–84

ELSEVIER

Contents lists available at ScienceDirect

Paediatric Respiratory Reviews

Review

New modes in non-invasive ventilation

Claudio Rabec [1,2,*], Guillaume Emeriaud [3], Alessandro Amadeo [4,5,6], Brigitte Fauroux [4,5,6], Marjolaine Georges [1,2]

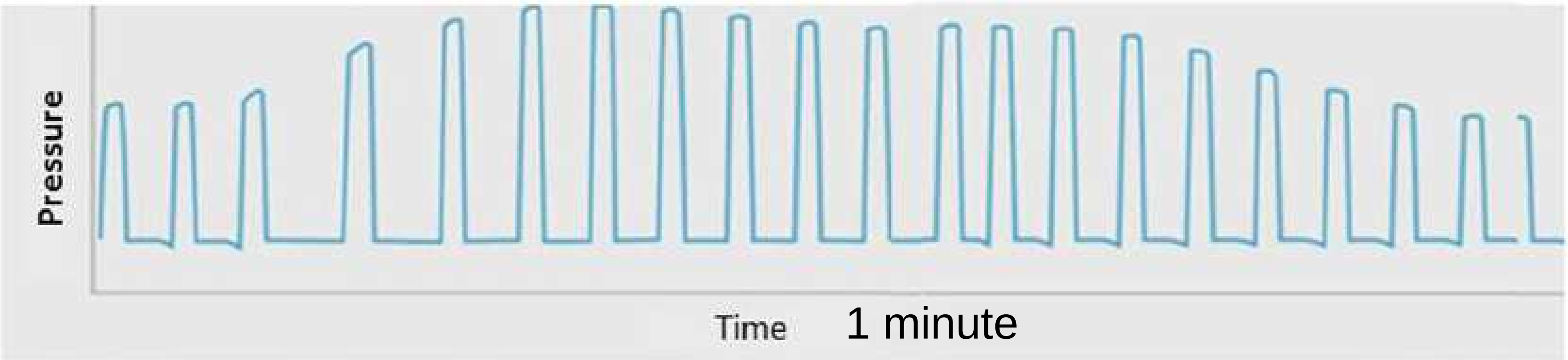
	Characteristics	Brands
Target volume with variable pressure support	Automatically adjust IPAP level (in a predefined pressure rang) to achieve a stable predetermined target Vt	AVAPS™ (A40™, Trilogy 100™ and 200™ *Philips*) Target volume pressure support (Vivo™ 50 and 60, *Breas*; Ventilogic™, *Weinmann*, Monnal T50™, *ALMS*; Elysee™ 150, 250, 350, *Resmed*)
Target volume with both variable pressure support and back-up respiratory rate	Automatically adjust both IPAP and BURR level (in a predefined pressure rang) to achieve a stable target predetermined minute ventilation	IVAPS™ (VPAP S9™, Stellar™ 100 and 150, Lumis™ Astral™, *Resmed*)

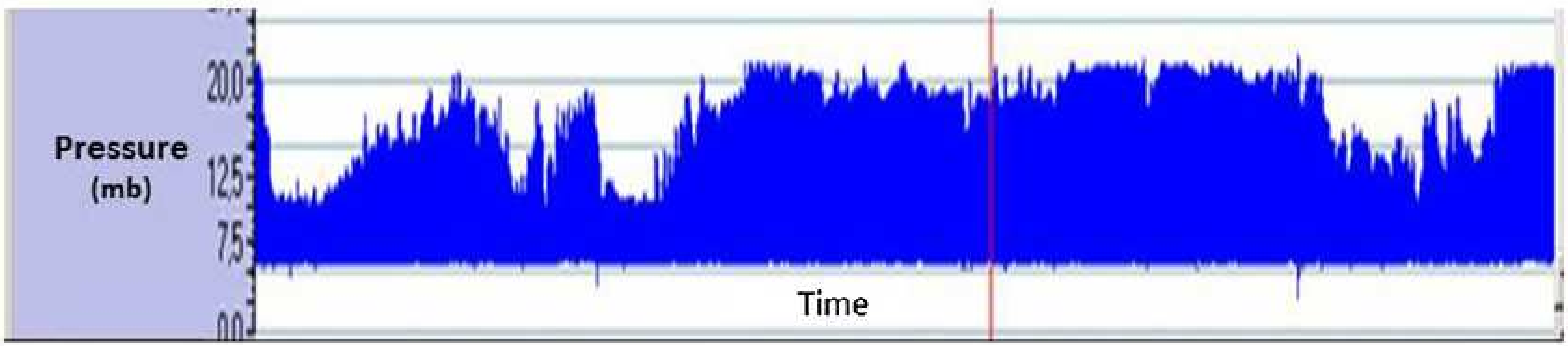
Target volume pressure support

IVAPS

AVAPS

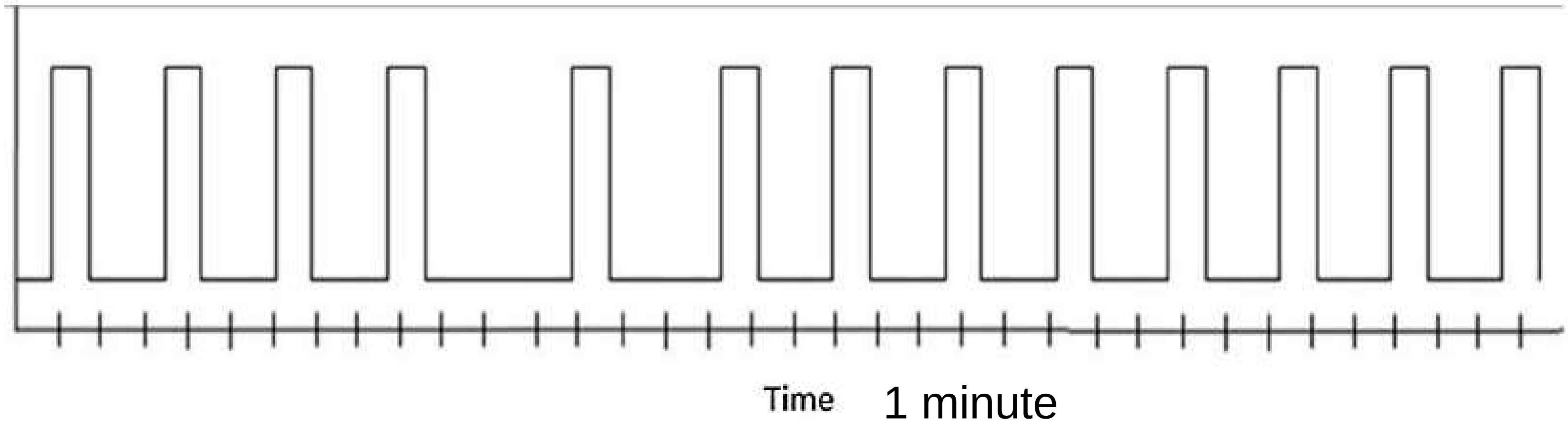
Target volume with variable pressure support



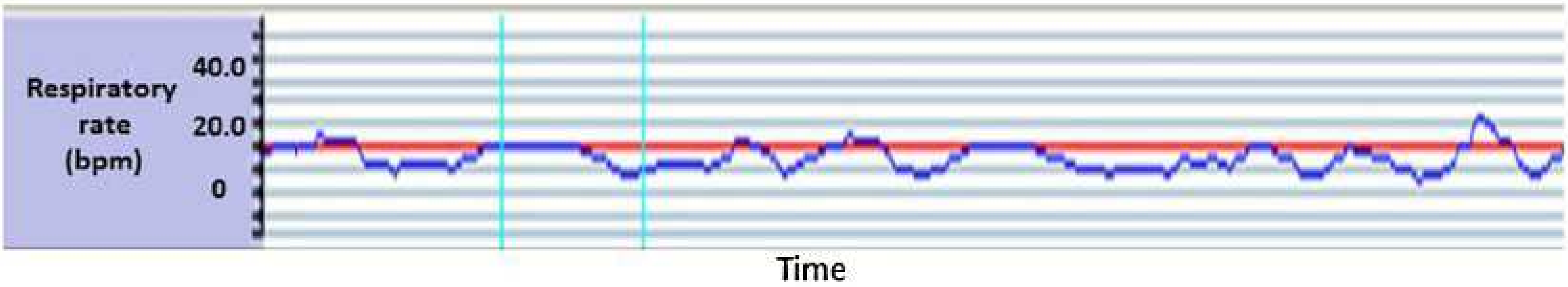


Entire night

Target volume with variable pressure support and back up rate



1 minute



Entire night

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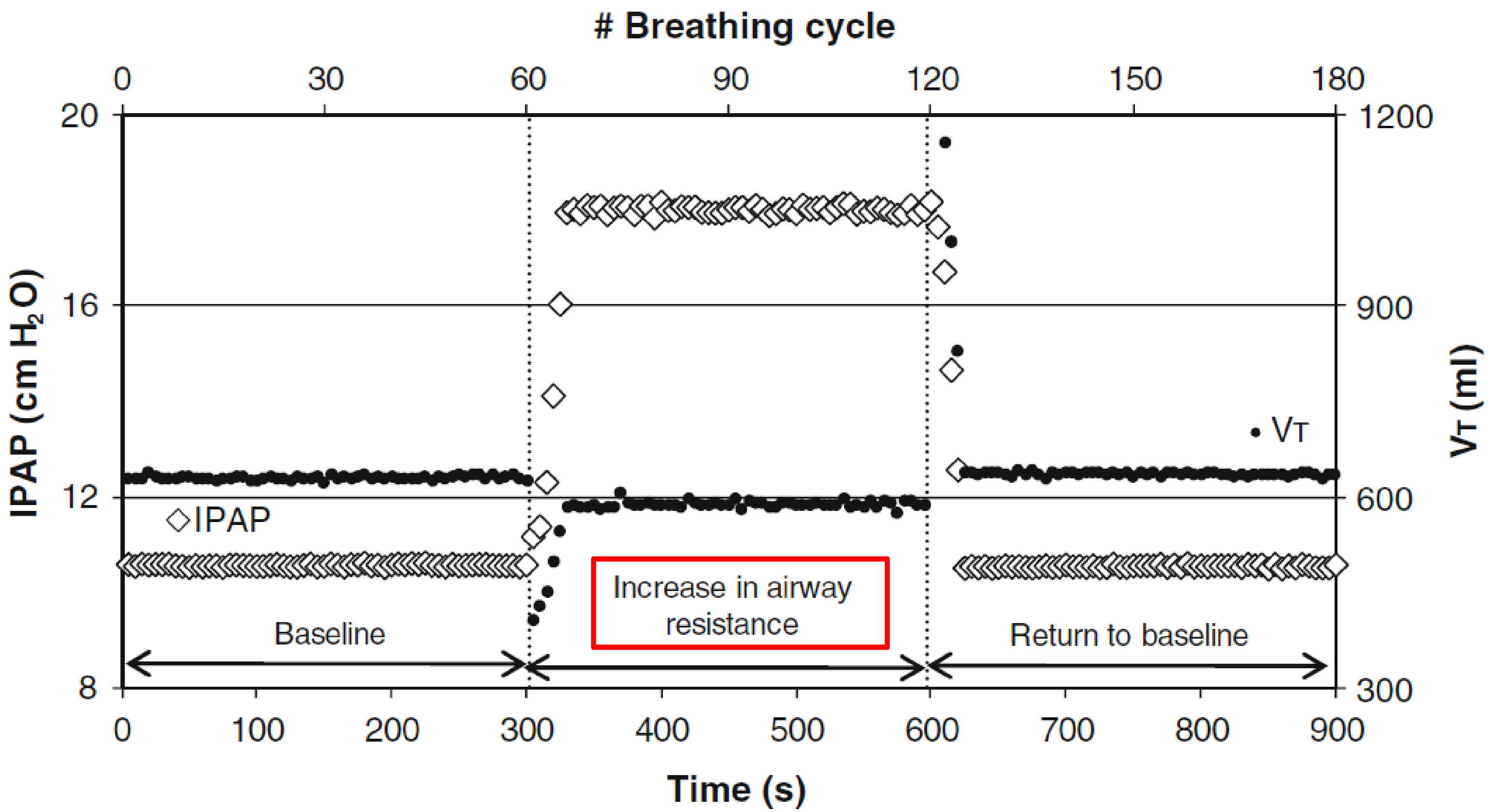
Intensive Care Med (2010) 36:1008–1014
DOI 10.1007/s00134-010-1785-9

ORIGINAL

Brigitte Fauroux
Karl Leroux
Jean-Louis Pépin
Frédéric Lofaso
Bruno Louis

Are home ventilators able to guarantee a minimal tidal volume?



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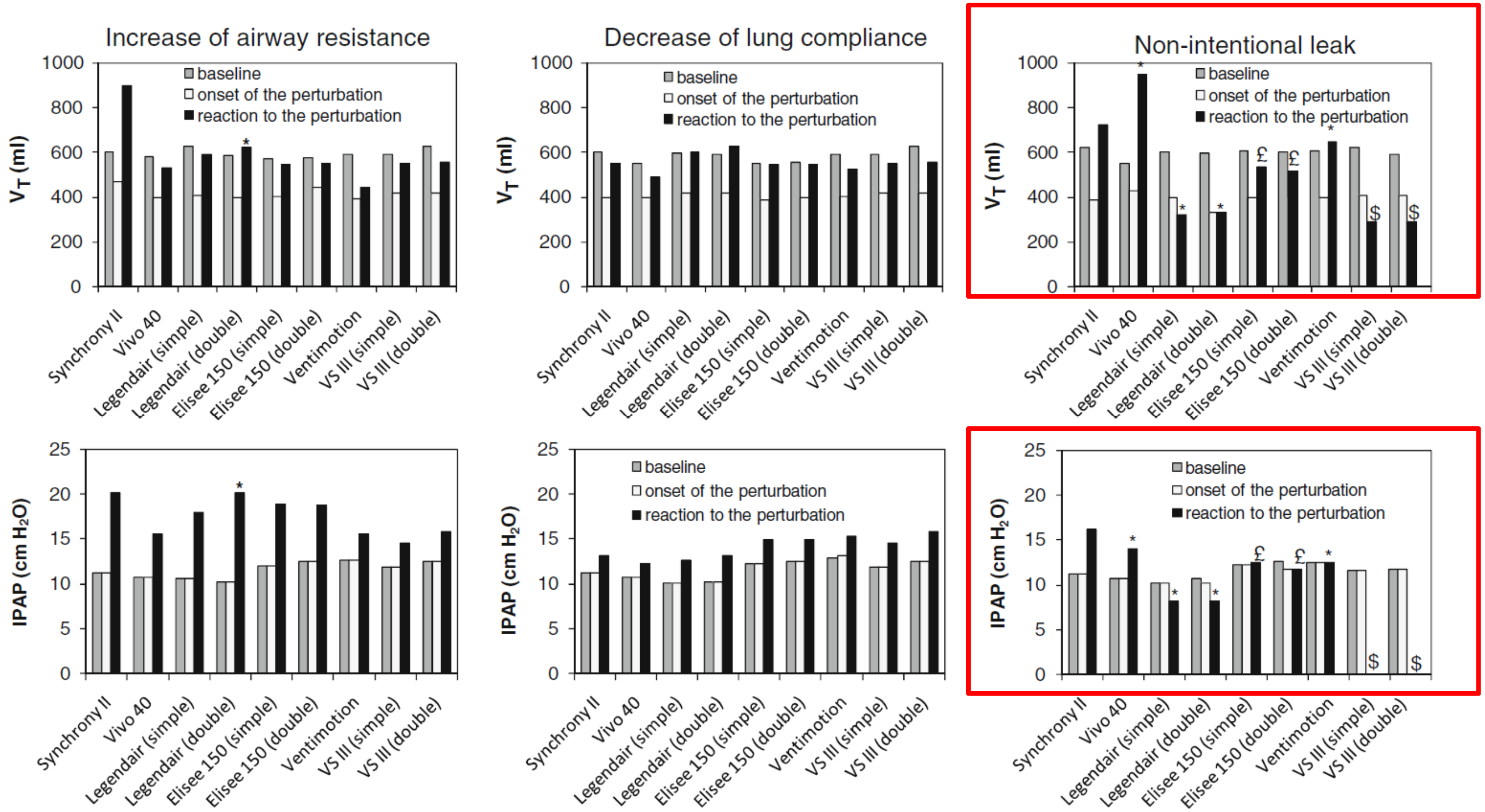

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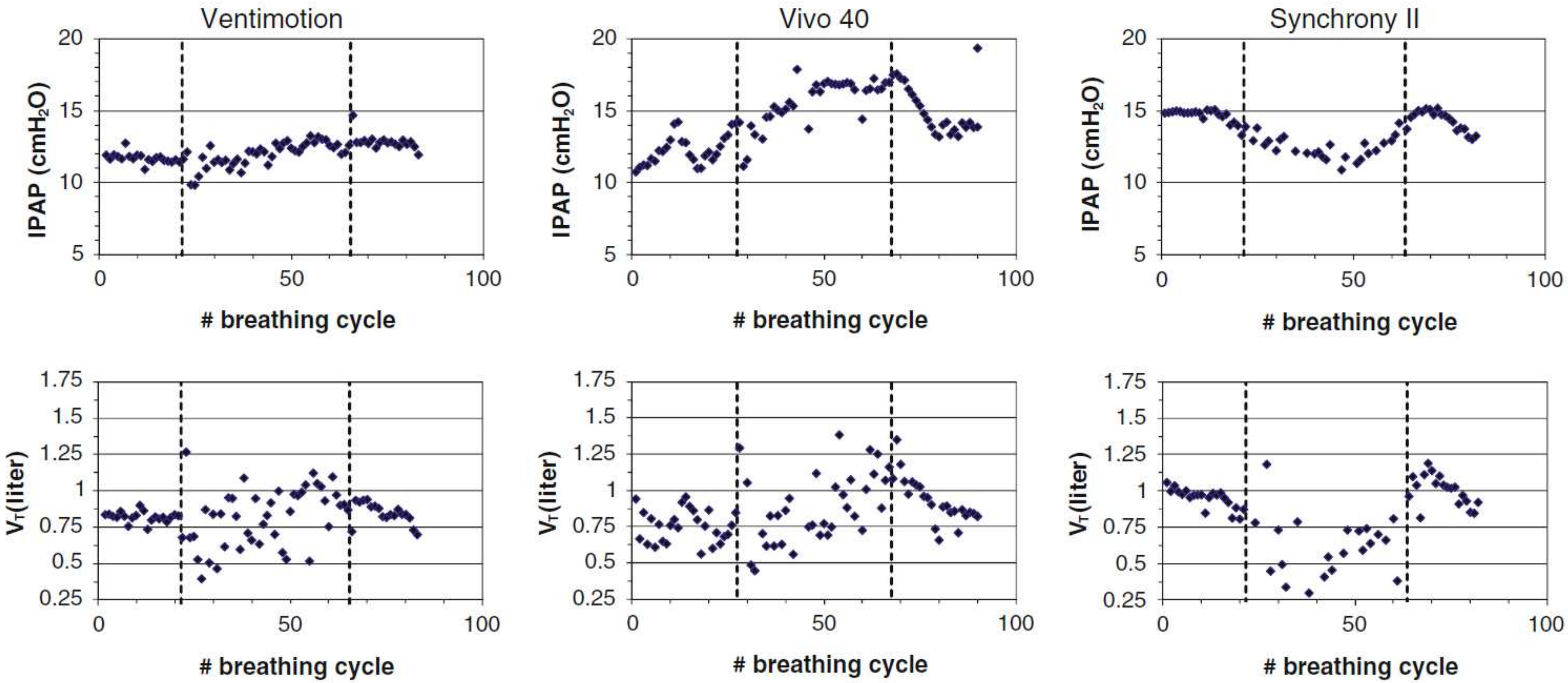
Are home ventilators able to guarantee a minimal tidal volume?

Evolution of IPAP and VT during mouth leaks



Harms of unintentional leaks during volume targeted pressure support ventilation

Sonia Khirani [a,b,c], Bruno Louis [c,d,e], Karl Leroux [f], Vincent Delord [b], Brigitte Fauroux [b,c], Frédéric Lofaso [c,d,e,g,h,*]

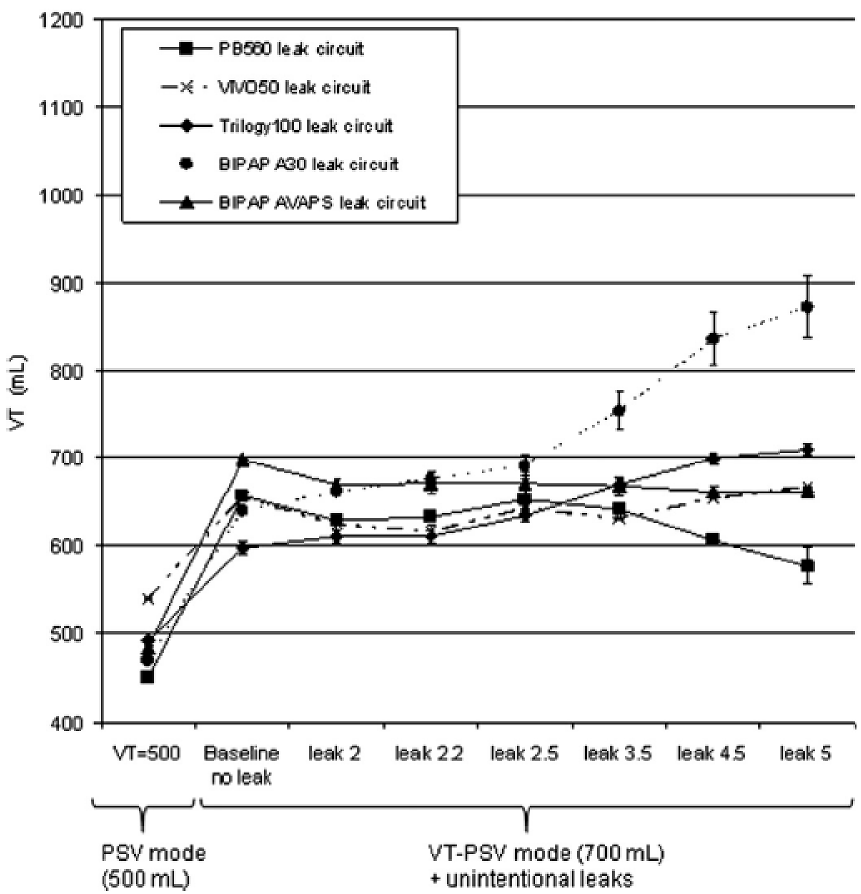
Table 1 Ventilatory settings and circuits.

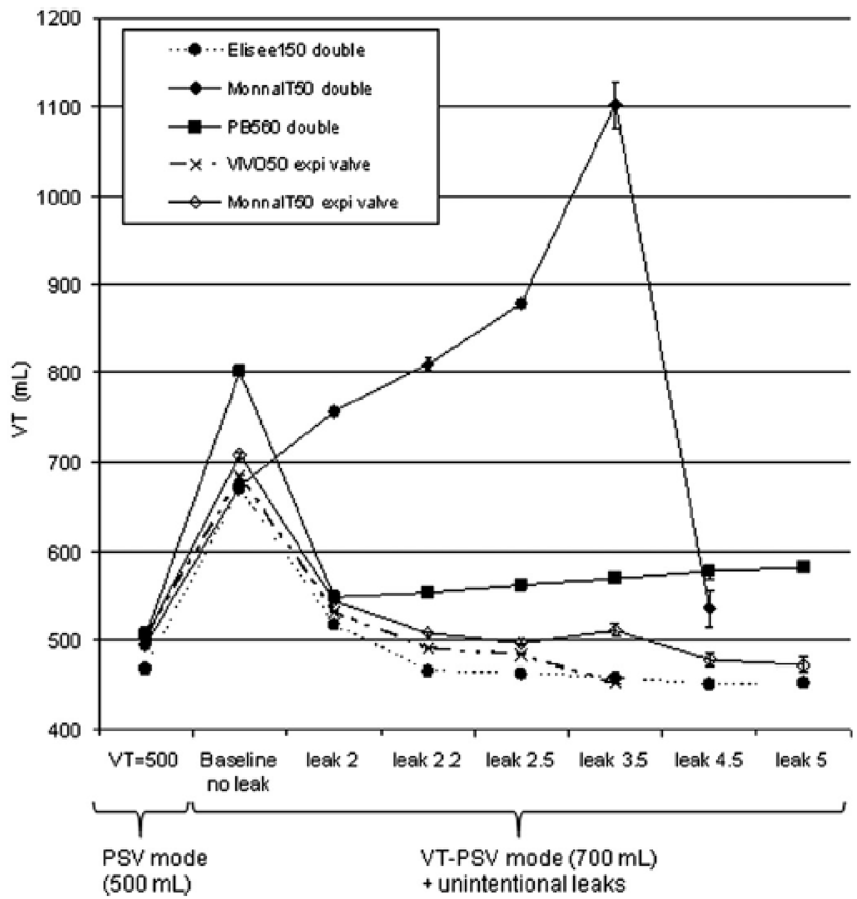
	Mode	Circuit	IPAPmin (cmH_2O)	IPAPmax (cmH_2O)	PEEP (cmH_2O)	F (b/min)	TI (s)	TRIG I (L/min)	TRIG E	Slope
ELISEE 150 (ResMed SA, Saint Priest, France)	AI VS	D	10		4	15	1.3	5	25%	1
MONNAL T50 (Air Liquide,	AI VS	S	10	34	4	15	1.3	10	30%	1
Antony, France)	AI VS	D	10	34	4	15	1.3	10	30%	1
PB560 (COVIDIEN Courtaboeuf,	AI FR	D	10	30	4	15	—	5	35%	1
France)	AI FR	S + leak	10	30	4	15	—	5	35%	1
VIVO 50 (Breas Medical	VS-Vtcibl	S	10	20	4	15	1.3	9	9 L/min	1
Saint Priest, France)	VS-Vtcibl	S + leak	11	20	4	15	1.3	9	9 L/min	1
TRILOGY 100 (Respironics France Carquefou, France)	ST-AVAPS	S + leak	10	34	4	15	1.3	9	30%	1
BIPAP A30 (Respironics France Carquefou, France)	ST-AVAPS	S + leak	10	25	4	15	1.3	Auto	Auto	1
BIPAP AVAPS (Respironics France Carquefou, France)	ST-AVAPS	S + leak	10	25	4	15	1.3	Auto	Auto	1

Respiratory Medicine (2013) **107**, 1021—1029

Harms of unintentional leaks during volume targeted pressure support ventilation

Sonia Khirani [a,b,c], Bruno Louis [c,d,e], Karl Leroux [f],
Vincent Delord [b], Brigitte Fauroux [b,c], Frédéric Lofaso [c,d,e,g,h,*]





Respiratory Medicine (2013) **107**, 1021—1029

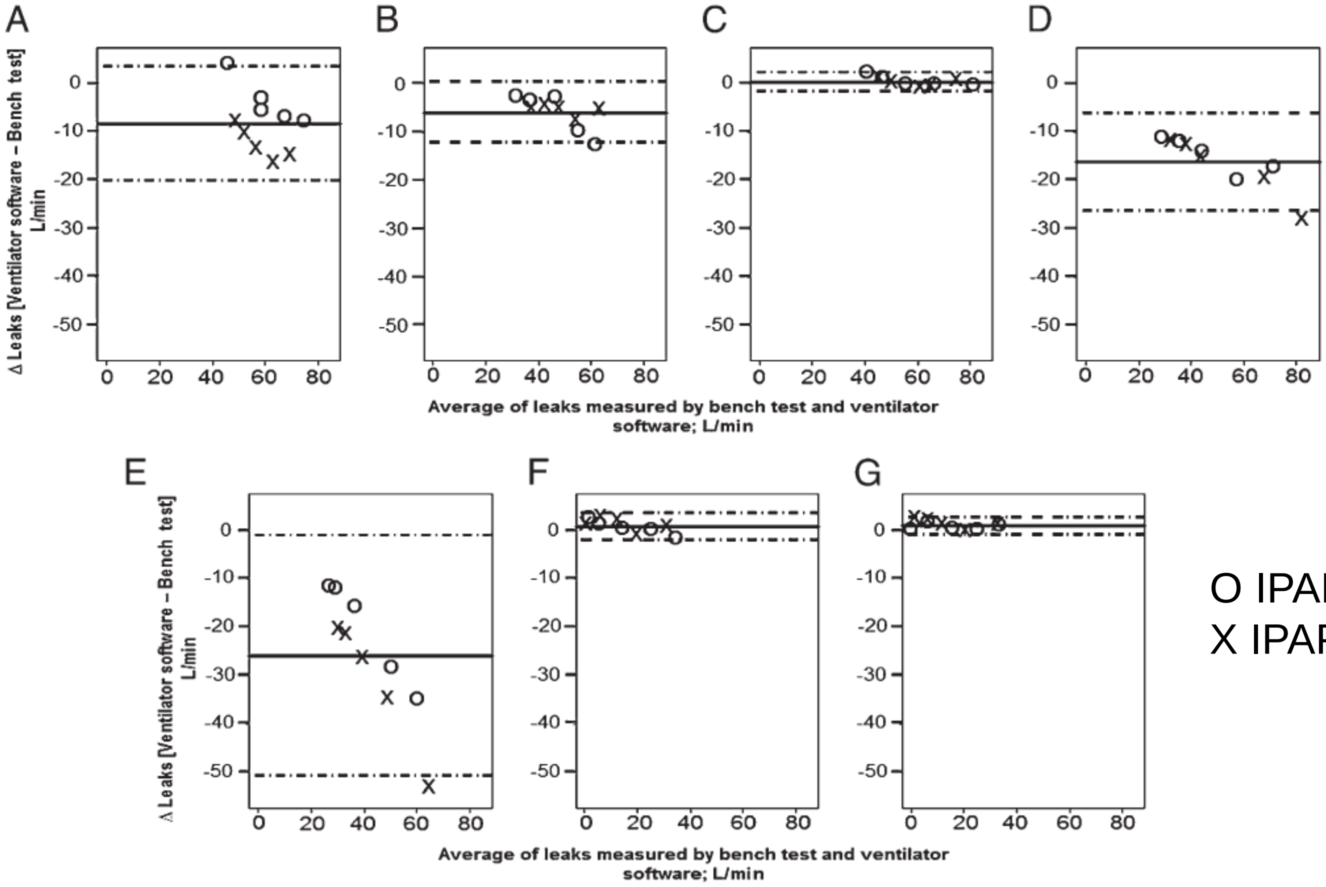
Monitoring of Noninvasive Ventilation by Built-in Software of Home Bilevel Ventilators

A Bench Study

CHEST 2012; 141(2):469–476

Olivier Contal, MS PT; Laurence Vignaux, MS PT; Christophe Combescure, PhD; Jean-Louis Pepin, MD, PhD; Philippe Jolliet, MD; and Jean-Paul Janssens, MD

Difference in leaks calculated on the bench and given by the in-built software



O IPAP 15 cmH_2O
X IPAP 25 cmH_2O

Monitoring of Noninvasive Ventilation by Built-in Software of Home Bilevel Ventilators

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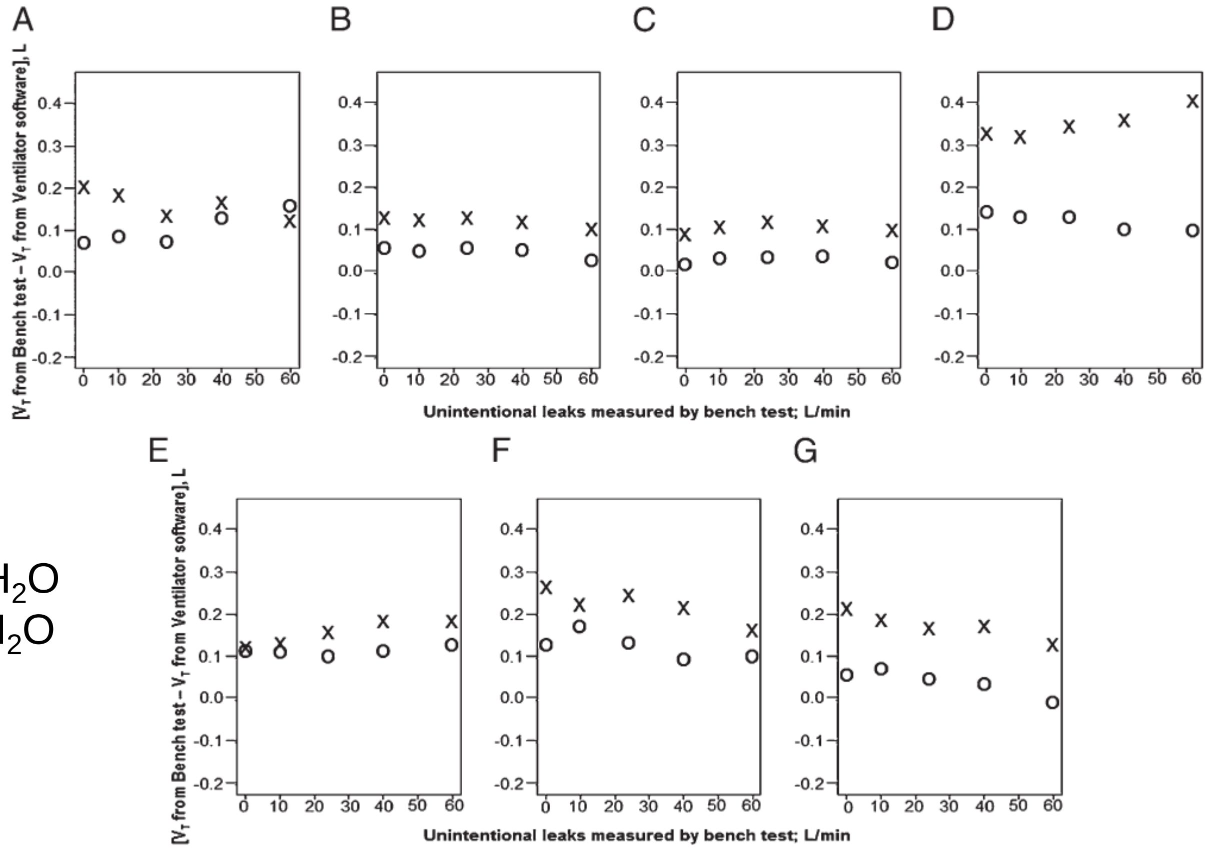
Olivier Contal, MS PT; Laurence Vignaux, MS PT; Christophe Combescure, PhD; Jean-Louis Pepin, MD, PhD; Philippe Jolliet, MD; and Jean-Paul Janssens, MD

Difference in tidal volume calculated on the bench and given by the in-built software



O IPAP 15 cmH_2O
X IPAP 25 cmH_2O

Pressure Support Versus Assisted Controlled Noninvasive Ventilation in Neuromuscular Disease

Karim Chadda,[1] Bernard Clair,[1] David Orlikowski,[1] Gilles Macadoux,[1] Jean Claude Raphael,[1] and Frédéric Lofaso[1,2,]*

13 adults with NMD requiring NIV, 3 modes in a random order, duration 25 min

Arterial Blood Gas and Respiratory Parameters During SB, ACV, APVC, and PSV

	SB	*ACV*	*APCV*	*PSV*	*ANOVA, p*
V_T, L	0.31 ± 0.12	0.71 ± 0.20[a]	0.74 ± 0.29[a]	0.77 ± 0.23[a]	<0.0001
RR, breaths/minute	21 ± 6	17 ± 2	16 ± 3	13 ± 3[a]	<0.0001
V_E, L/minute	6.0 ± 1.8	12.2 ± 3.3[a]	11.8 ± 5.0[a]	9.7 ± 1.8	<0.0001
T_I, s	1.42 ± 0.45	1.37 ± 0.24	1.41 ± 0.28	1.64 ± 0.32	0.08
V_T/T_I, L/second	0.23 ± 0.06	0.54 ± 0.20[a]	0.54 ± 0.26[a]	0.48 ± 0.13[a]	<0.0001
Vi max, L/second	0.30 ± 0.10	0.59 ± 0.19	0.76 ± 0.24	0.83 ± 0.19[b]	<0.0001
$PaCO_2$, mmHg	51± 8	43 ± 8	45 ± 13	43 ± 7	<0.005
PaO_2, mmHg	74 ± 10	80 ± 11	84 ± 11	84 ± 12	<0.005
pH	7.39± 0.05	7.43 ± 0.05	7.43 ± 0.06	7.44 ± 0.05	<0.005

Abbreviations: SB, spontaneous breathing; ACV, assisted controlled ventilation; APCV, assisted pressure-controlled ventilation; PSV, pressure-support ventilation; V_T, inspiratory tidal volume; RR, respiratory rate; V_E, minute ventilation; T_I, inspiratory time; V_T/T_I, mean inspiratory flow; Vi max, maximal inspiratory flow.

Pressure Support Versus Assisted Controlled Noninvasive Ventilation in Neuromuscular Disease

Karim Chadda,[1] Bernard Clair,[1] David Orlikowski,[1] Gilles Macadoux,[1] Jean Claude Raphael,[1] and Frédéric Lofaso[1,2,]*

Mechanical and Respiratory Effort Parameters During SB, ACV, APVC, and PSV

	SB	*ACV*	*APCV*	*PSV*	*ANOVA, p*
$PEEPI_{dyn}$, cm H_2O	0.21± 0.19	0.23 ± 0.19	0.28 ± 0.16	0.23 ± 0.19	NS
CL_{dyn}, L/cm H_2O	0.088 ± 0.074	0.088 ± 0.041	0.088 ± 0.037	0.079 ± 0.047	NS
Swing Pes, cm H_2O	6.39 ± 2.10	1.54 ± 1.02[a]	1.67 ± 1.00[a]	2.10 ± 1.05[a]	<0.0001
Swing Pdi, cm H_2O	7.52 ± 4.12	1.31 ± 1.01[a]	1.36 ± 1.09[a]	2.11 ± 1.11[a]	<0.0001
PTPes, cm H_2O.s/minute	126 ± 35	54 ± 45[a]	62 ± 65[a]	70 ± 42[a]	<0.0001
PTPdi, cm H_2O.s/minute	161 ± 74	59 ± 61[a]	64 ± 52[a]	66 ± 40[a]	<0.0001
Cycle Triggering, %	—	4.5 ± 12.3	14.5 ± 29.4	33.6 ± 26.9[b]	0.007

			Triggering %		
Patient no.	*Patient choice*	*<PTPes*	*ACV*	*APCV*	*PSV*
1	PSV	APCV	10	90	50
2	PSV	ACV	0	0	20
3	ACV	ACV	0	0	0
4	PSV	ACV	0	0	10
5	ACV	APCV	0	50	20
6	APCV	PSV	0	0	20
7	APCV	ACV	0	0	0
8	PSV	ACV	20	20	50
9	APCV	ACV	0	0	70
10	ACV	ACV	40	20	60
11	ACV	PSV	0	0	0
12	PSV	APCV	0	0	20
13	APCV	APCV	0	0	50

Randomized trial of 'intelligent' autotitrating ventilation versus standard pressure support non-invasive ventilation: Impact on adherence and physiological outcomes

JULIA L. KELLY,* JAY JAYE,* RACHEL E. PICKERSGILL, MICHELLE CHATWIN, MARY J. MORRELL AND ANITA K. SIMONDS *Respirology* (2014) **19**, 596–603

7/18 patients NM

Table 2 Ventilator output and adherence to therapy following treatment with iVAPS and standard PS non-invasive ventilation

	iVAPS	Standard PS	Median difference between treatments (95% CI)	*P*
Ventilator settings (*n* = 18)				
PS minimum and maximum boundaries (iVAPS) (cmH_2O)	5.0 (5.0–5.0)–17.5 (15.0–18.0)	n/a	n/a	n/a
PS (standard PS) (cmH_2O)	n/a	10.0 (9.0–11.4)	n/a	n/a
EPAP (cmH_2O)	7.8 (6.0–9.0)	7.3 (6.0–9.0)	0 (0 to 1)	0.77
RR (bpm)	16.5 (14.0–21.0)	12.0 (12.0–13.0)	4.7 (2.3 to 7.3)	0.001†
Target Va (l/min)	4.9 (4.1–6.1)	n/a	n/a	n/a
Ventilator output (*n* = 16)				
PS delivered median (cmH_2O)‡	8.3 (5.6–10.4)	10.0 (9.0–11.4)	−2.2 (−4.5 to 0.3)	0.001†
Median leak (l/min)—vent	6.5 (3.5–26)	3.6 (0.2–9.6)	3.5 (−2.5 to 9.6)	0.23
Median tidal volume (mL)	421 (321–521)	400 (300–575)	−10 (−54 to 23)	0.47
Median minute ventilation (l/min)	6.8 (5.3–8.3)	6.2 (5.4–9.4)	−0.2 (−1.2 to 0.5)	0.50
Median RR (bpm)	16.7 (13.2–18.4)	15.5 (13.5–17.0)	0.3 (−0.7 to 2.2)	0.41
Adherence (*n* = 17)				
Mean NIV usage time (hh:mm/day)	5:40 (4:42–6:49)	4:20 (2:27–6:17)	01:04 (00:27 to 1:44)	0.004†
% days used in study	91 (64–98)	92 (70–99)	−1 (−15 to 7)	0.53
% days used ≥4/24	74 (49–92)	60 (27–85)	8 (−2 to 17)	0.1

No difference on PSG and nocturnal gas exchange

Intensive Care Med
DOI 10.1007/s00134-009-1408-5

ORIGINAL

Rebeca Paiva
Uros Krivec
Guillaume Aubertin
Emmanuelle Cohen
Annick Clément
Brigitte Fauroux

Carbon dioxide monitoring during long-term noninvasive respiratory support in children

Table 1 Characteristics of the patients

	Patients $n = 50$
Age (years)	8.5 ± 5.2
Male/Female	29/21
Primary disease	
Neuromuscular disease	23
Lung disease	2
Upper airway obstruction	25
Duration of noninvasive respiratory support (months)	24 ± 20
Ventilatory mode	
AC/VT	21
PS	19
CPAP	10
Nasal mask	
Industrial	25
Custom made	25

Intensive Care Med
DOI 10.1007/s00134-009-1408-5

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Table 2 Results of the nocturnal recording by the combined $PtcCO_2/SpO_2$ monitor

	Normal $PtcCO_2$ recording	Abnormal $PtcCO_2$ recording	Total number of patients $N = 50$
SpO_2 cut off of 90%			
$SpO_2 > 90\%$	28 (56%)	21 (42%)	49 (98%)
$SpO_2 \leq 90\%$	1 (2%)	0 (0%)	1 (2%)
SpO_2 cut off of 92%			
$SpO_2 > 92\%$	28 (46%)	18 (36%)	46 (92%)
$SpO_2 \leq 92\%$	3 (6%)	1 (2%)	4 (8%)
SpO_2 cut off of 95%			
$SpO_2 > 95\%$	18 (36%)	12 (24%)	30 (60%)
$SpO_2 \leq 95\%$	13 (26%)	7 (14%)	20 (40%)

Normal daytime blood gases and normal nocturnal SpO_2 do not exclude nocturnal hypercapnia

Daytime $PaCO_2$	Patients with nocturnal $PtcCO_2$ < 50 mmHg n=29 (%)	Patients with nocturnal $PtcCO_2$ > 50 mmHg n=21 (%)
$PaCO_2$ < 45 mmHg	29 (48%)	18 (36%)
$PaCO_2$ ≥ 45 mmHg	0 (0%)	3 (6%)

Paiva *et al.* Intensive Care Med 2009:35:1068

Intensive Care Med
DOI 10.1007/s00134-009-1408-5

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Table 4 Interventions (not exclusive) in the patients who had nocturnal hypercapnia during noninvasive positive pressure ventilation (NPPV)

	Patients, $n = 21$
Change of the settings of noninvasive respiratory support	7
Change of interface	6
Addition of a chin strap	4
Addition of an abdominal girdle	3
Change of NPPV mode	2
Adaptation of brace	2
Other intervention*	3
No change	3
Tracheostomy	1

Nocturnal Oximetry and Transcutaneous Carbon Dioxide in Home-Ventilated Neuromuscular Patients

Julie Nardi MD, Hélène Prigent MD, Annie Adala, Mikaëlle Bohic, François Lebargy MD PhD, Maria-Antonia Quera-Salva MD PhD, David Orlikowski MD PhD, and Frédéric Lofaso MD PhD

	Normal P_{tcCO_2}, no.*	Abnormal P_{tcCO_2}, no.*	Total, no.
S_{pO_2} criterion 1			
Normal	33	22	55
Abnormal	1	2	3
Total	34	24	58
S_{pO_2} criterion 2			
Normal	33	17	50
Abnormal	1	7	8
Total	34	24	58
S_{pO_2} criterion 3			
Normal	30	15	45
Abnormal	4	9	13
Total	34	24	58

Detection of alveolar hypoventilation by SpO_2 or with $PtcCO_2$:

- $SpO_2 \leq 88\% > 5$min
- Mean $SpO_2 < 90\%$ or $SpO_2 < 90\% \geq 10\%$ recording time
- Mean $SpO_2 < 92\%$ or $SpO_2 < 90\% \geq 10\%$ recording time
- $PtcCO_2$ max ≥ 49mmHg

	Diagnosis of Alveolar Hypoventilation, no. (%)
S_{pO_2} criterion 1	3 (5.2)
S_{pO_2} criterion 2	8 (13.8)
S_{pO_2} criterion 3	13 (22.4)
P_{tcCO_2}	24 (41.4)
P_{tcCO_2} or S_{pO_2} criterion 2	25 (43.1)
P_{tcCO_2} or S_{pO_2} criterion 3	28 (48.3)

No effect of ventilatory mode

Respiratory Care • September 2012 Vol 57 No 9 frontiers in Medicine

ORIGINAL RESEARCH
published: 13 September 2016

Prognostic Value of Initial Assessment of Residual Hypoventilation Using Nocturnal Capnography in Mechanically Ventilated Neuromuscular Patients: A 5-Year Follow-up Study

Adam Ogna[1], Julie Nardi[2], Helene Prigent[2], Maria-Antonia Quera Salva[3], Cendrine Chaffaut[4], Laure Lamothe[1], Sylvie Chevret[4], Djillali Annane[1], David Orlikowski[1,5] and Frederic Lofaso[2,3]*

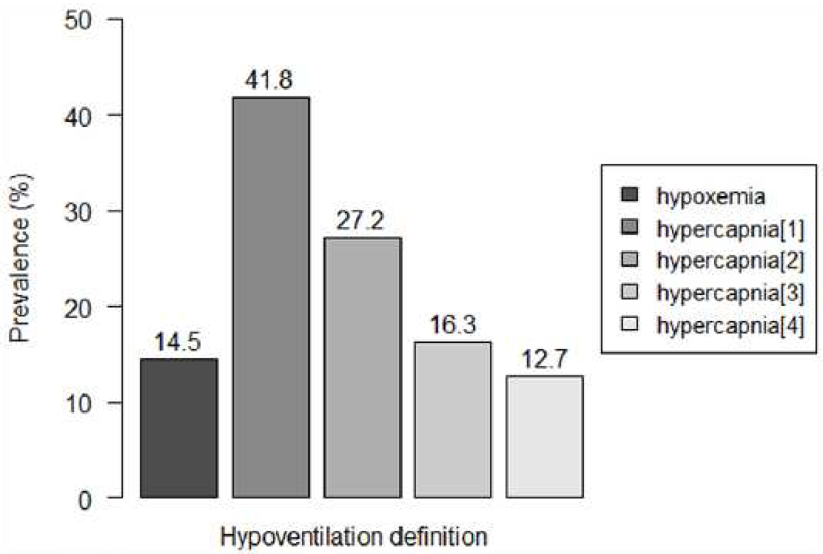
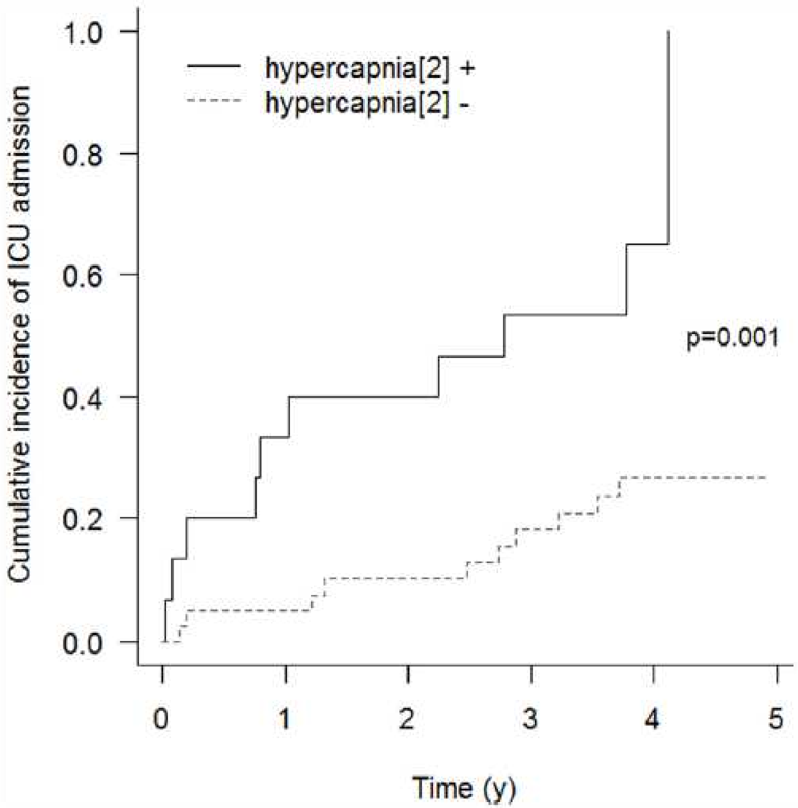


TABLE 1 | Characteristics of the study population.

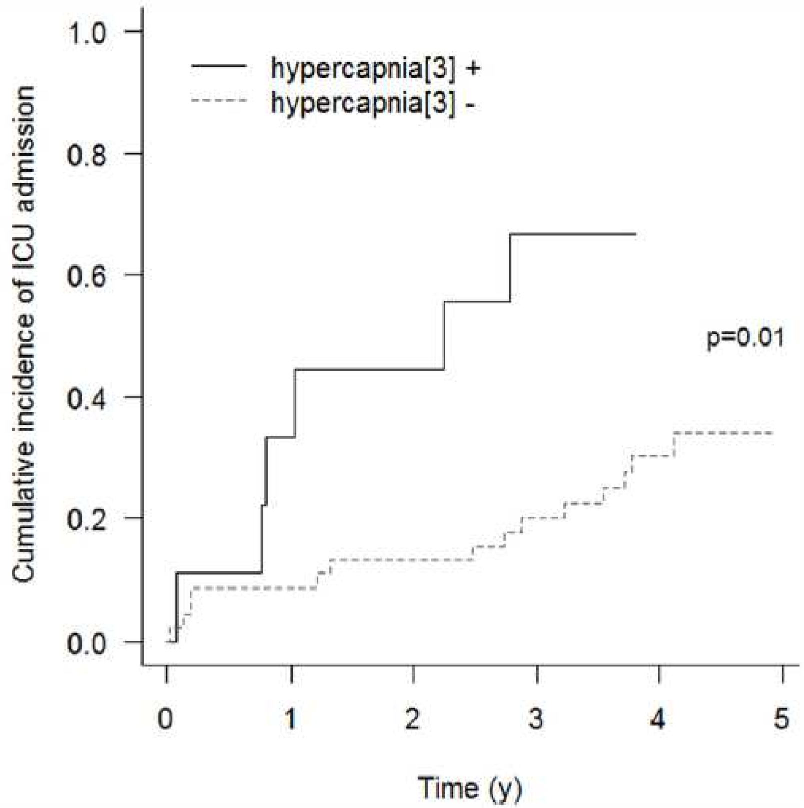
	N (%) or median [IQR]
Parameters	
Number of patients	55
Pathology (*N*, %)	
– DMD	39 (70.9%)
– MD1	5 (9.1%)
– Other	11 (20.0%)
Age (years)	28 [25–36.5]
Weight (kg)	45.0 [36.0–60.5]
BMI (kg/m^2)	17.0 [14.6–24.2]
Follow-up (years)	4.0 [3.6–4.5]
Deaths (*N*, %)	12 (21.8%)
ICU admissions (*N*, %)	20 (36.4%)
Respiratory parameters	
VC sitting (%pred)	12 [7–27]
VC supine (%pred)	10 [5–20]
PI max (cmH_2O)	12 [3–27]
PE max (cmH_2O)	10 [5–24]
Mechanical ventilation	
Volumetric mode (*N*, %)	40 (72.7%)
Tracheostomy (*N*, %)	28 (50.9%)
Daily HMV duration (h)	22.5 [9.0–24.0]

Cumulative incidence of respiratory events requiring ICU admission

$PtcCO_2$ > 49 mmHg > 10% total recording time

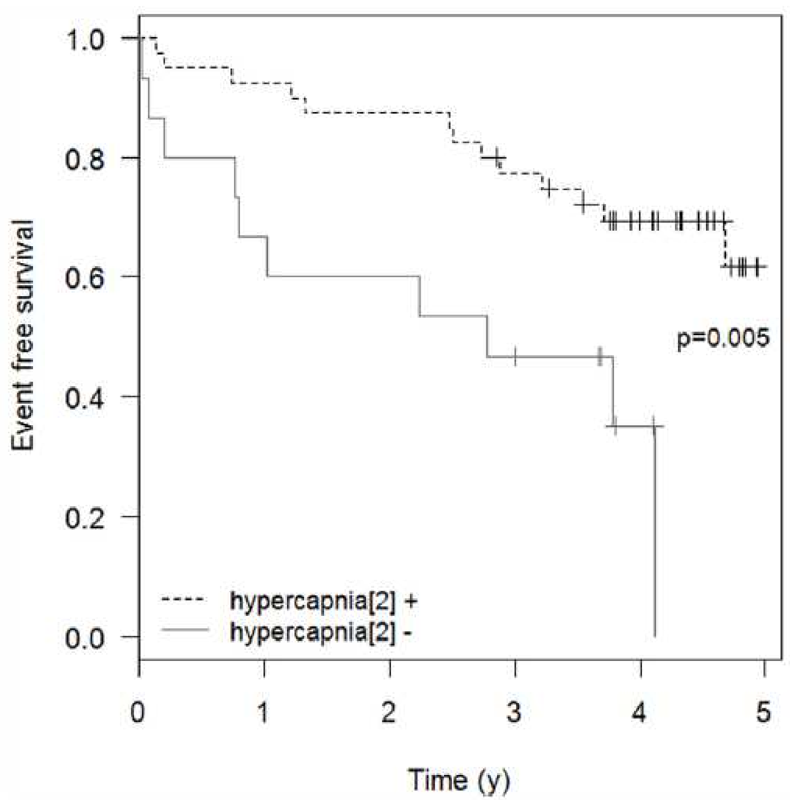


peak $PtcCO_2$ > 55 Hg



Event free survival: time to ICU admission or death

PtcO$_2$ > 49 mmHg > 10% total recording time



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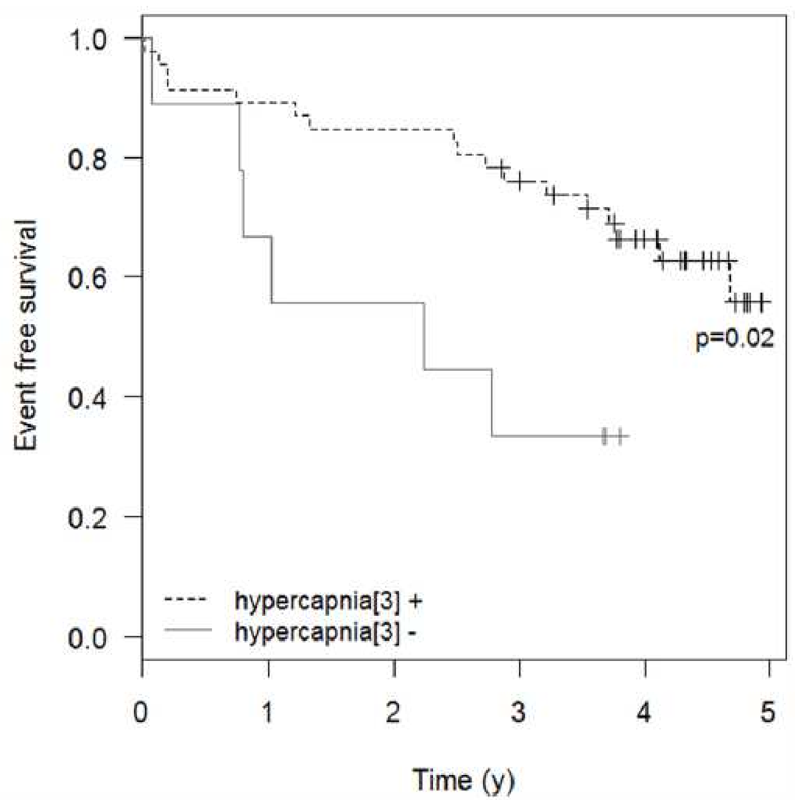

Which ventilatory mode for children with neuromuscular diseases ?

- Aim of NIV: guarantee an adequat (physiological) tidal volume with an optimal comfort
- NM patients are « easy » to ventilate: NIV replaces the respiratory muscles
  - adequat inspiratory trigger (sensitive) or back up rate = physiological breathing rate
  - expiratory pressure = 0 ou minimal
- No ventilatory mode has proven its superiority
- In practice: S/T + volume guarantee + back up rate

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PEDIATRIC ORIGINAL

Valeria Caldarelli
Jean Christian Borel
Sonia Khirani
Adriana Ramirez
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Jean-Louis Pépin
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Polygraphic respiratory events during sleep with noninvasive ventilation in children: description, prevalence, and clinical consequences

Table 2 Description of the respiratory events during a polygraphy recording: percentage of recording time spent with a single respiratory event (mean ± SD) and number of subjects with the single events

	Unintentional leaks	Patient–ventilator asynchronies	Decrease in ventilatory drive	Upper airway obstruction with decrease in ventilatory drive	Upper airway obstruction	Mixed events
NM disease ($n = 13$)	31 ± 38 % ($n = 7$)	38 ± 46 % ($n = 9$)	8 ± 16 % ($n = 6$)	11 ± 23 % ($n = 5$)	4 ± 7 % ($n = 6$)	0 ± 0 % ($n = 0$)
Lung disease ($n = 11$)	37 ± 41 % ($n = 8$)	50 ± 39 % ($n = 10$)	4 ± 8 % ($n = 5$)	8 ± 12 % ($n = 5$)	16 ± 18 % ($n = 7$)	0.1 ± 0.1 % ($n = 1$)
OSAS ($n = 15$)	17 ± 31 % ($n = 6$)	11 ± 24 % ($n = 6$)	17 ± 33 % ($n = 10$)	20 ± 34 % ($n = 7$)	16 ± 25 % ($n = 9$)	9 ± 26 % ($n = 8$)
Total population ($n = 39$)	27 ± 36 % ($n = 21$)	33 ± 40 % ($n = 25$)	10 ± 23 % ($n = 21$)	11 ± 28 % ($n = 17$)	12 ± 10 % ($n = 22$)	3 ± 16 % ($n = 9$)

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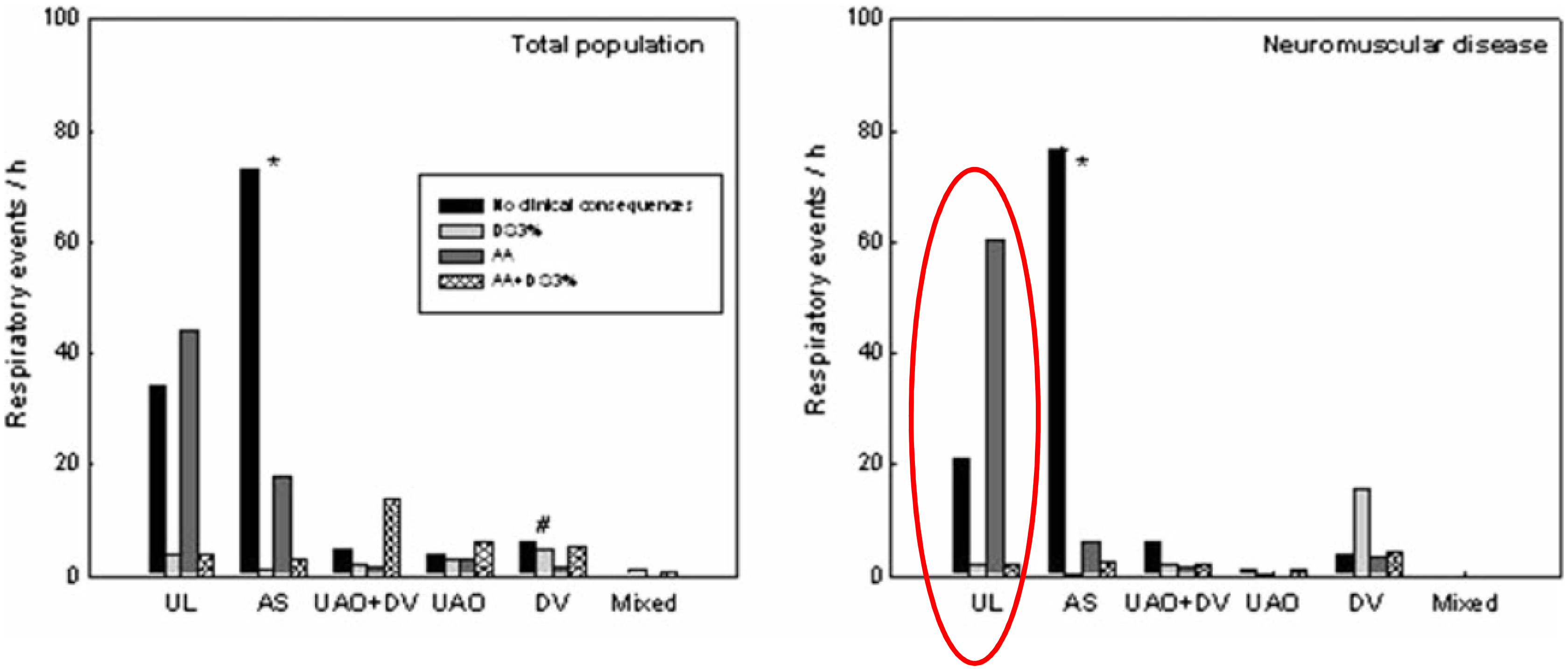

Non intentional leaks are associated with autonomic arousals

Conclusion

- Outbreak of « new » ventilatory modes
  - lack of harmonisation of definitions ++
  - none has proven its superiority
- Primary aim
  - guarantee a physiological tidal volume with maximal comfort
- Priorities > ventilatory mode
  - for the patient
    - interface and headgear ++
    - ergonomy of the ventilator: weight, encumbrance, humidification
    - possibility of different modes (mouthpiece ventilation)
  - for the medical staff
    - easiness and accuracy of in-built software
    - integrated SpO_2 monitoring ($PtcCO_2$?)

Acknowledgements

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