3rd INTERNATIONAL PEDIATRIC NONINVASIVE VENTILATION CONFERENCE

Necker university hospital
Paris - France
November 7th & 8th 2019

Initiation of NIV: for which children, when and how ?

Pr Brigitte Fauroux

Pediatric noninvasive ventilation and sleep unit
Paris Descartes University EA 7330 VIFASOM
(Vigilance Fatigue Sommeil et Santé Publique)
Necker university hospital, Paris, France

Université de Paris

Necker ENFANTS MALADES HÔPITAL UNIVERSITAIRE

IRBA

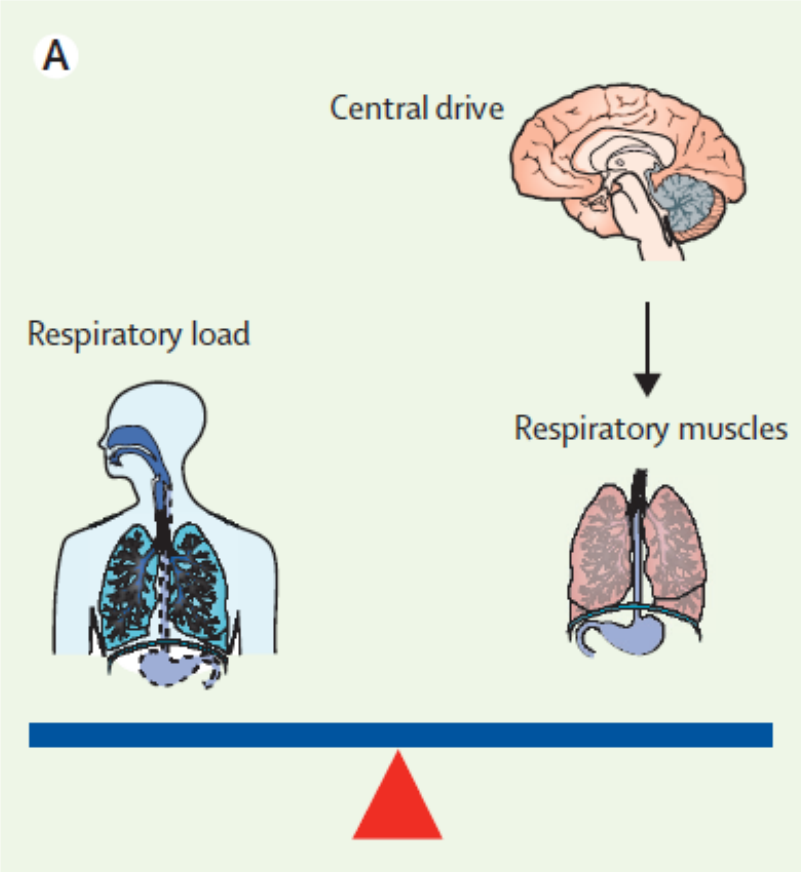
antadir INDÉPENDANCE ET EXPERTISE POUR LE PROGRÈS DE LA SANTÉ À DOMICILE

NIV in Pediatrics

- Which patients ?
- When to start ?
- Where to start ?
- Follow up and weaning ‘

Indications

Normal respiratory balance



Long-term non-invasive ventilation in children

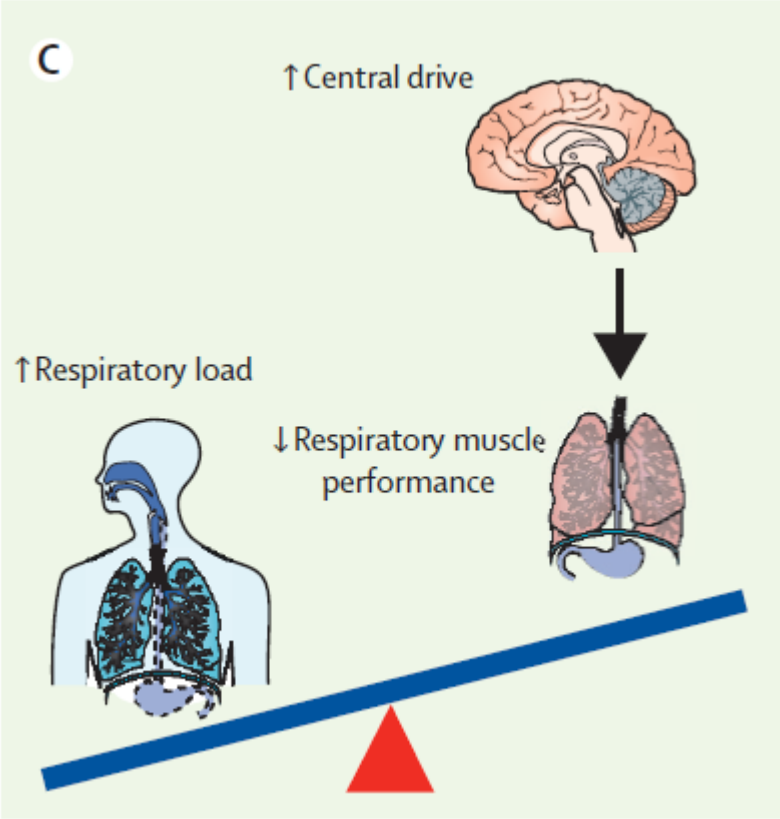
Alessandro Amaddeo, Annick Frapin, Brigitte Fauroux

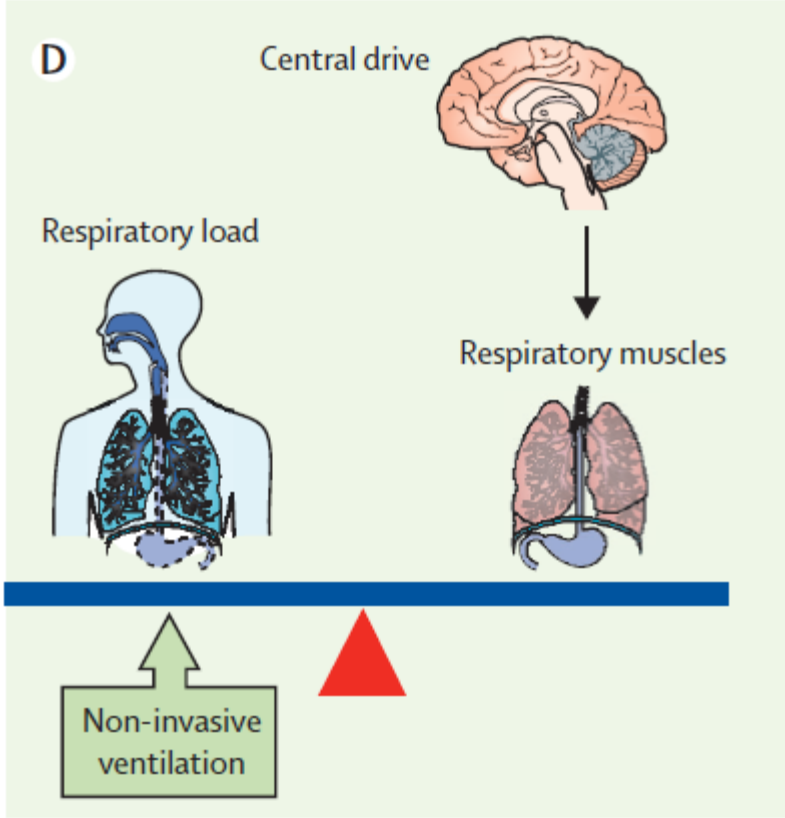
Lancet Respir Med 2016

Published Online
July 13, 2016

Indications

Disruption of the respiratory balance





Long-term non-invasive ventilation in children

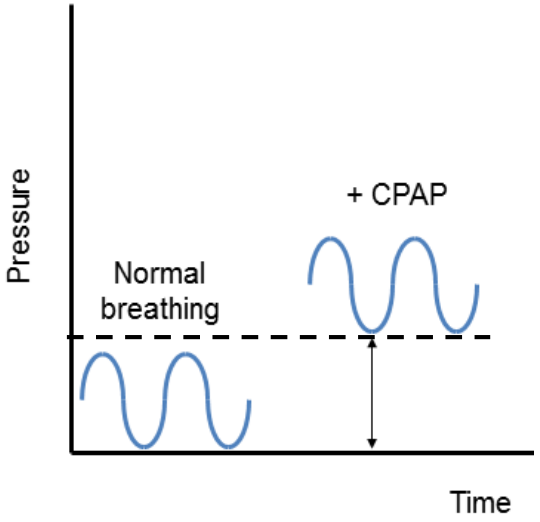
Alessandro Amaddeo, Annick Frapin, Brigitte Fauroux

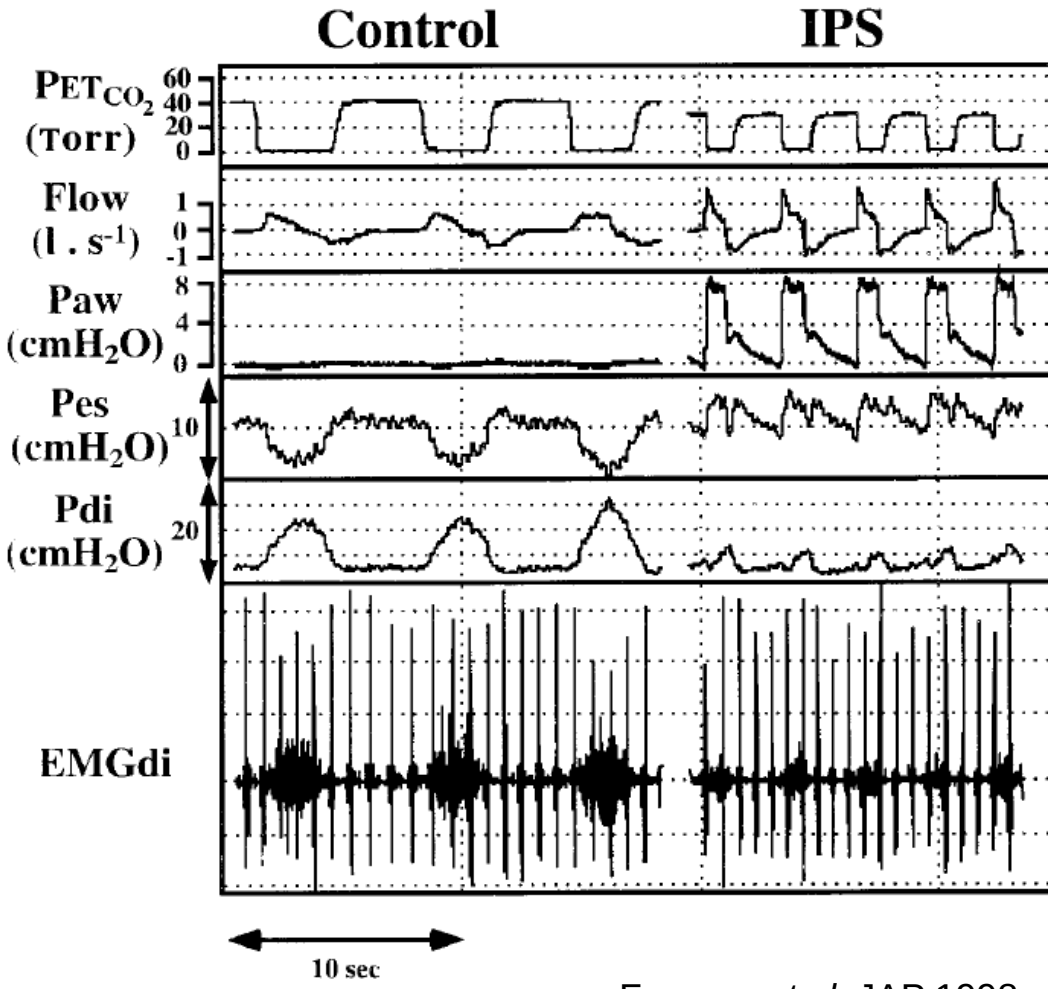
Lancet Respir Med 2016

Published Online
July 13, 2016

CPAP and NIV

CPAP





Fauroux *et al.* JAP 1998

Long Term Non-Invasive Ventilation in Children: Impact on Survival and Transition to Adult Care

PLOS ONE

May 1, 2015

Michelle Chatwin[1,3]*, Hui-Leng Tan[2,3], Andrew Bush[2,3], Mark Rosenthal[2,3], Anita Kay Simonds[1,3]

Table 2. Patients under the age of 17 who were commenced and ceased NIV and CPAP by group diagnosis.

Diagnosis Group	Total patients% (n)	Total patients onNIV% (n)	Total patientson CPAP% (n)
Chest wall disease	4 (16)	100 (16)	0 (0)
Obesity	3 (12)	75 (9)	25 (3)
Chronic lung disease	5 (24)	87 (21)	13 (3)
Central sleep apnoea	3 (12)	100 (12)	0 (0)
Cardiac surgery	2 (11)	36 (4)	64 (7)
Congenital syndrome	14 (61)	56 (34)	44 (27)
Upper airway abnormality	9 (39)	62 (24)	38 (15)
Other	4 (20)	95 (19)	5 (1)
Neuromuscular disease	56 (254)	96.6 (253)	0.4 (1)

496 children on long term NIV followed at the Royal Brompton hospital between 1993-2011

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]

144 children on long term NIV followed
over a 15-yr period in Vancouver, Canada

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)

Arch Dis Child 2013;**98**:660–665.

Longitudinal changes in clinical characteristics and outcomes for children using long-term non-invasive ventilation

PLOS ONE

January 30, 2018

Maria L. Castro-Codesal[1,2,3]*, Kristie Dehaan[1], Prabhjot K. Bedi[1], Glenda N. Bendiak[4,5], Leah Schmalz[5], Sherri L. Katz[6,7], Joanna E. MacLean[1,2,3]

Retrospective analysis of 622 children < 18 yrs on long term NIV between 2005 and 2014 in Alberta, Canada

Table 5. Longitudinal trends in the technology for children using long-term non-invasive ventilation.

	Jan 2005-Apr 2008 (n = 127)	May 2008-Aug 2011 (n = 262)	Sept 2011-Dec 2014 (n = 233)	*P* Value
NIV type [a]; n, %				
CPAP	101, 80 (95%CI 72–86)	207, 79 (95%CI 74–84)	171, 73 (95%CI 67–78)	0.19
BPAP	25, 20 (95%CI 14–28)	52, 20 (95%CI 15–25)	62, 26 (95%CI 21–32)	0.17
Auto-PAP	1, <1 (95%CI 0.1–4)	3, 1 (95%CI 0.4–3)	2, <1 (95%CI 0.2–3)	>0.99

Indications

Increase in respiratory load

Anatomical abnormalities of the upper airway

- Treacher Collins syndrome
- Craniofaciostenosis
- Pierre Robin syndrome
- Pycnodysostosis
- Achondroplasia
- Tracheomalacia or laryngomalacia
- Congenital or acquired laryngotracheal stenosis
- Vocal cord paralysis
- Other upper airway malformation
- Storage diseases
- Neck masses or tumours
- Down's syndrome
- Beckwith-Wiedemann syndrome

CPAP

Lower airway obstruction

- Cystic fibrosis
- Bronchopulmonary dysplasia
- Bronchiolitis obliterans

CPAP or NIV

Decrease in performance of respiratory muscles

- Spinal muscular atrophy
- Spinal cord injury
- Phrenic nerve injury
- Myasthenia
- Myopathies and dystrophies

NIV

Dysfunction of central drive

- Congenital central hypoventilation syndrome
- Brain injury by tumours or infection (encephalitis)
- Brainstem dysfunction (eg, Chiari malformation)

+ obesity/hypoventilation

Long-term non-invasive ventilation in children

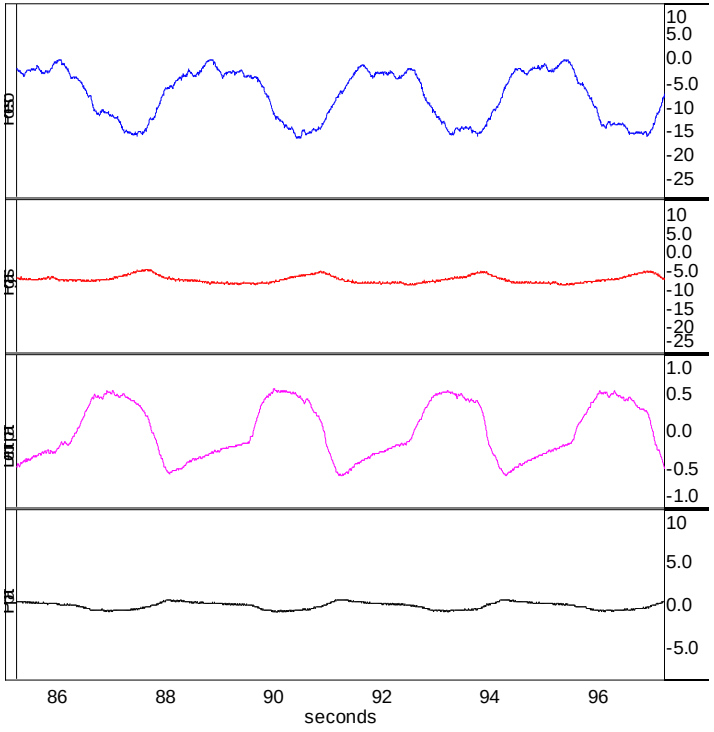
Alessandro Amaddeo, Annick Frapin, Brigitte Fauroux

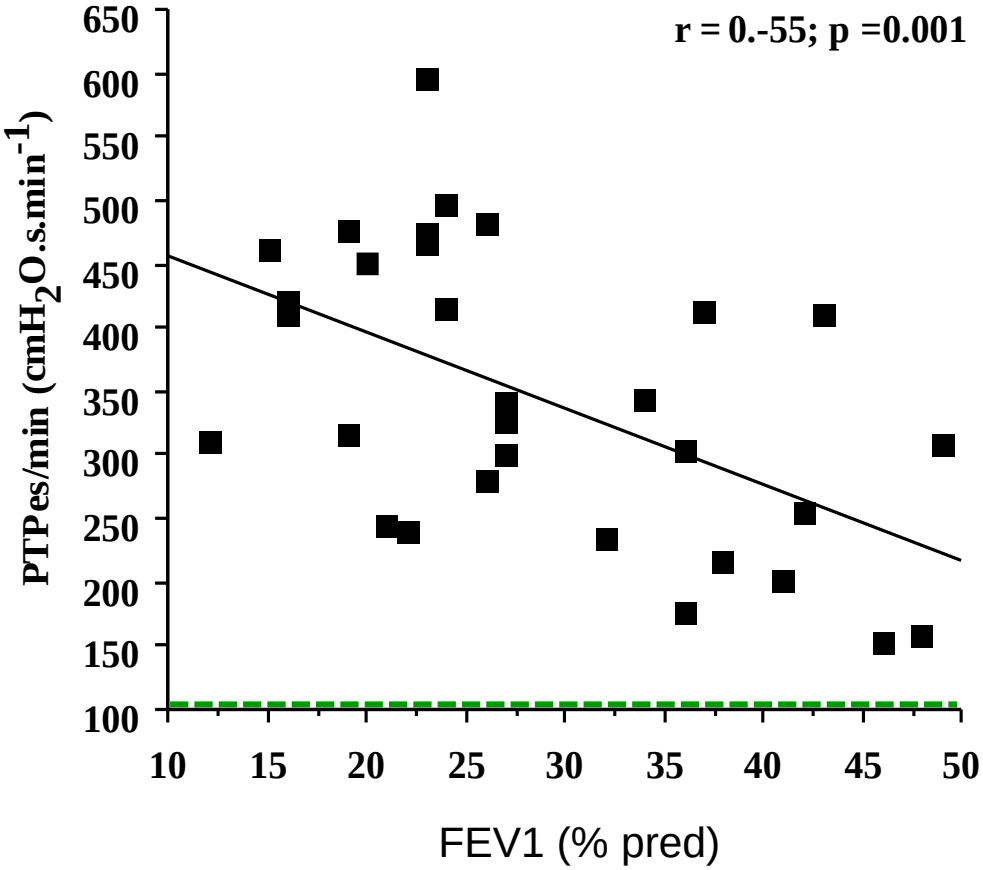
Lancet Respir Med 2016

Published Online
July 13, 2016

Changes in Pulmonary Mechanics with Increasing Disease Severity in Children and Young Adults with Cystic Fibrosis

Nicholas Hart, Michael I. Polkey, Annick Clément, Michèle Boulé, John Moxham, Frédéric Lofaso, and Brigitte Fauroux

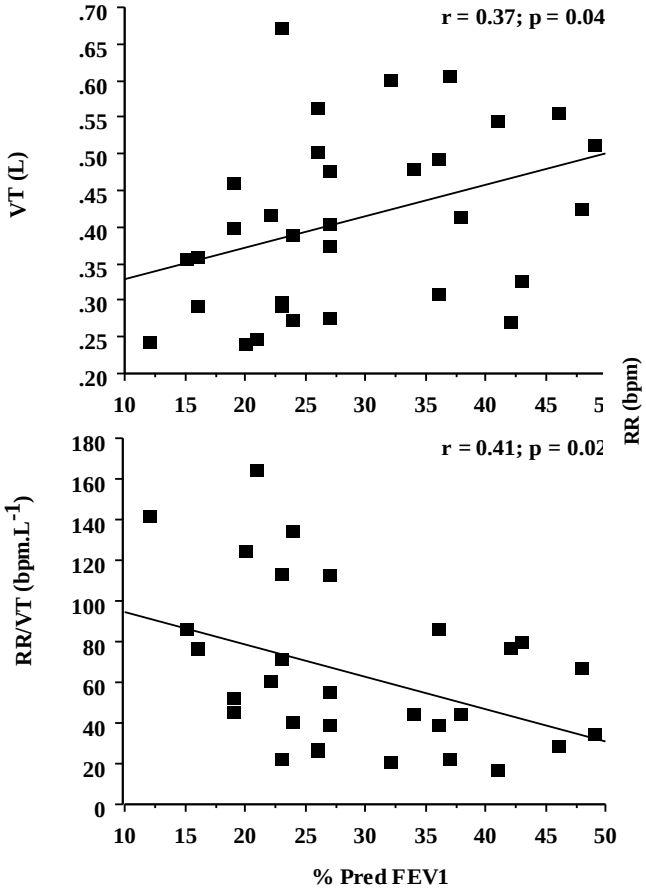


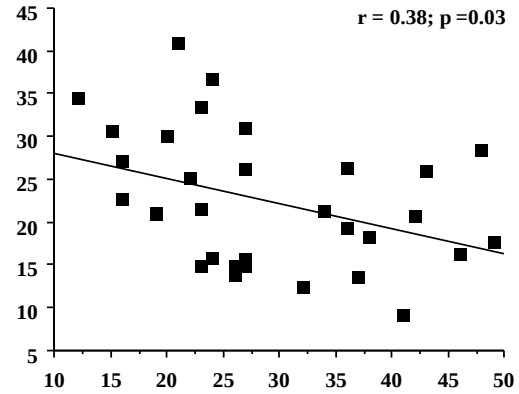


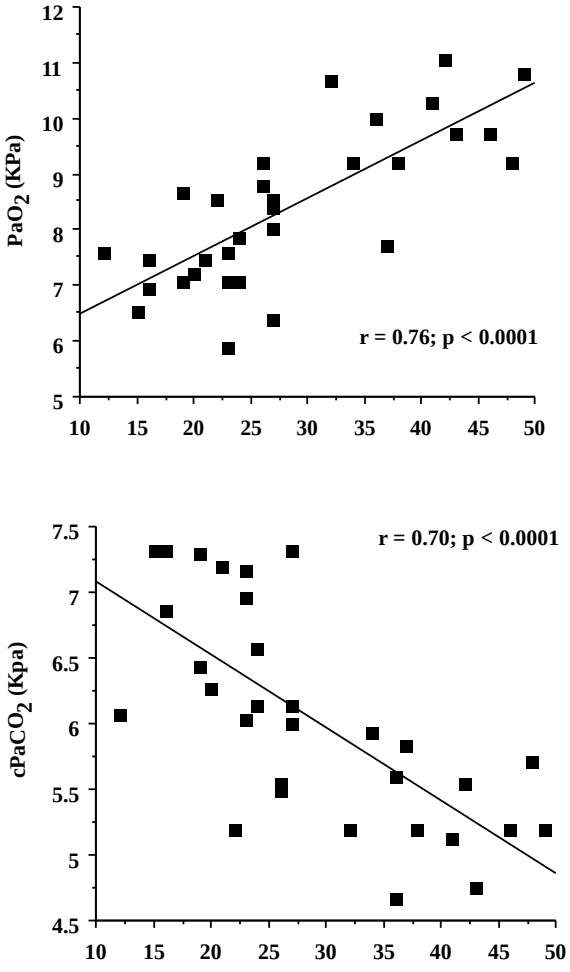
Am J Respir Crit Care Med Vol 166. pp 61–66, 2002

Changes in Pulmonary Mechanics with Increasing Disease Severity in Children and Young Adults with Cystic Fibrosis

Nicholas Hart, Michael I. Polkey, Annick Clément, Michèle Boulé, John Moxham, Frédéric Lofaso, and Brigitte Fauroux



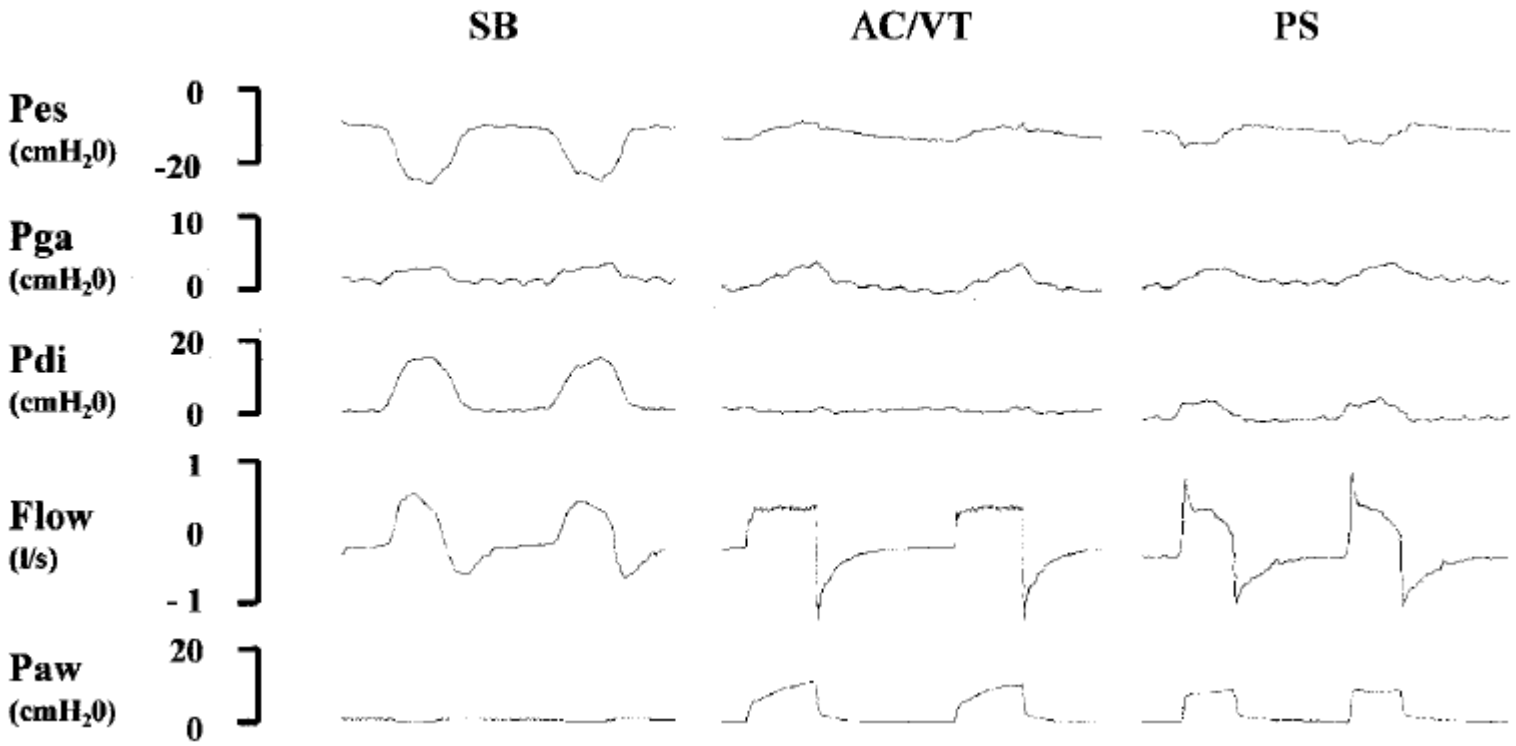




Am J Respir Crit Care Med Vol 166. pp 61–66, 2002

In vivo physiologic comparison of two ventilators used for domiciliary ventilation in children with cystic fibrosis

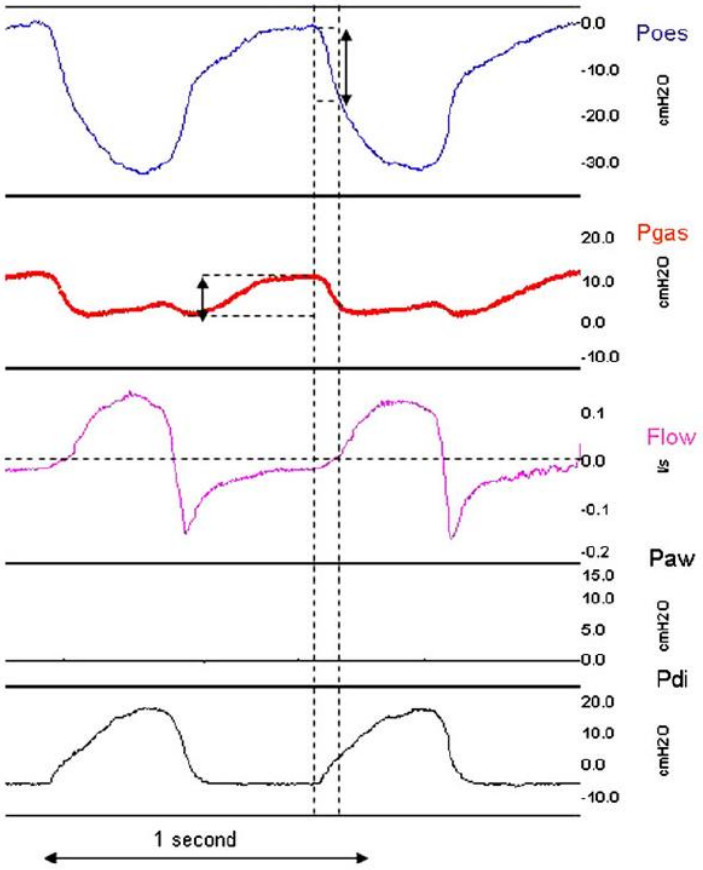
Brigitte Fauroux, MD, PhD; Jérôme Pigeot, MsC; Michael I. Polkey, MRCP, PhD; Daniel Isabey, PhD; Annick Clément, MD, PhD; Frédéric Lofaso, MD, PhD

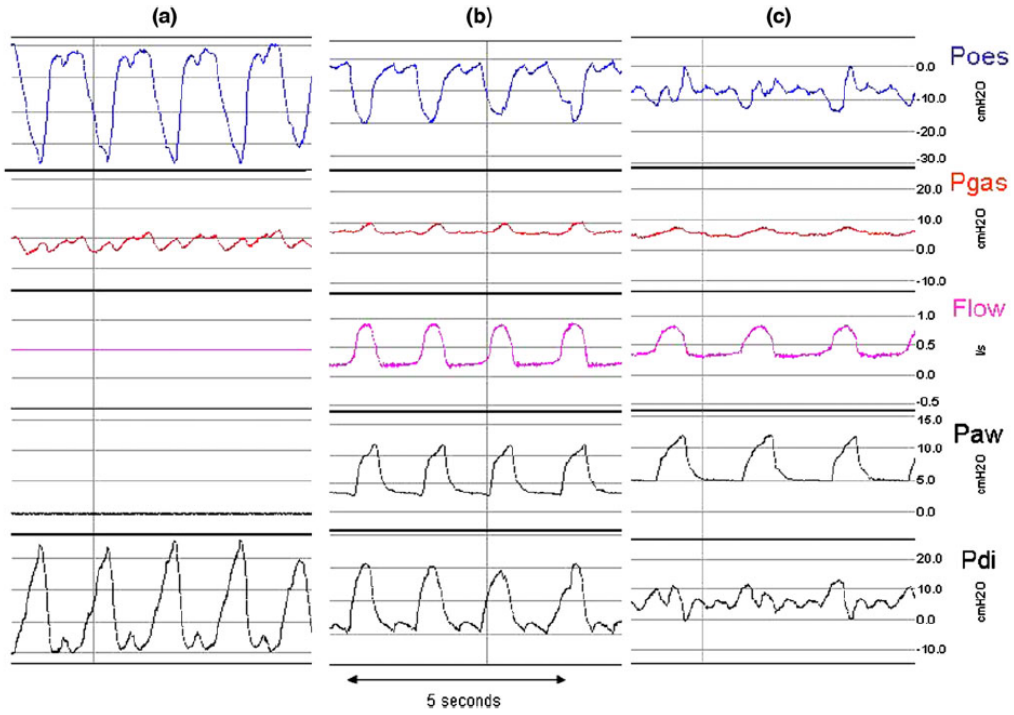


Crit Care Med 2001; 29:2097–2105

Lisa Giovannini-Chami
Sonia Khirani
Guillaume Thouvenin
Adriana Ramirez
Brigitte Fauroux

Work of breathing to optimize noninvasive ventilation in bronchiolitis obliterans



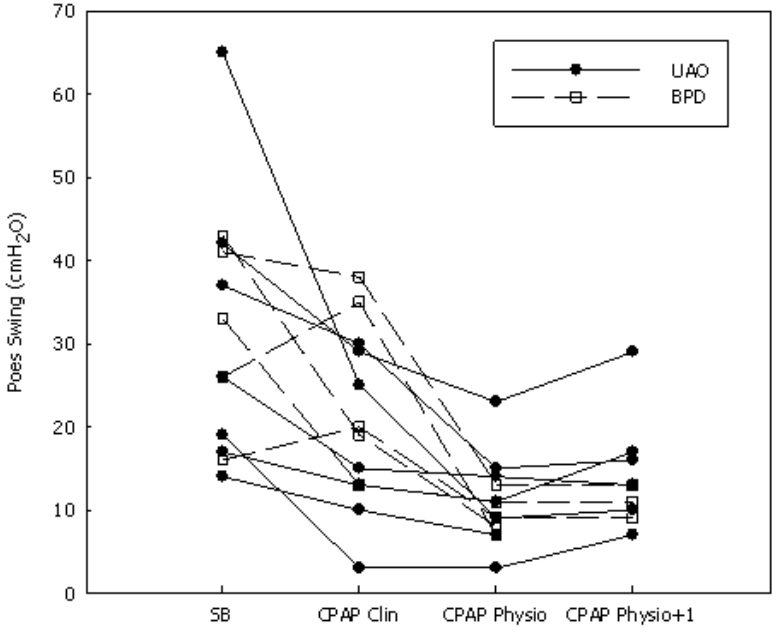


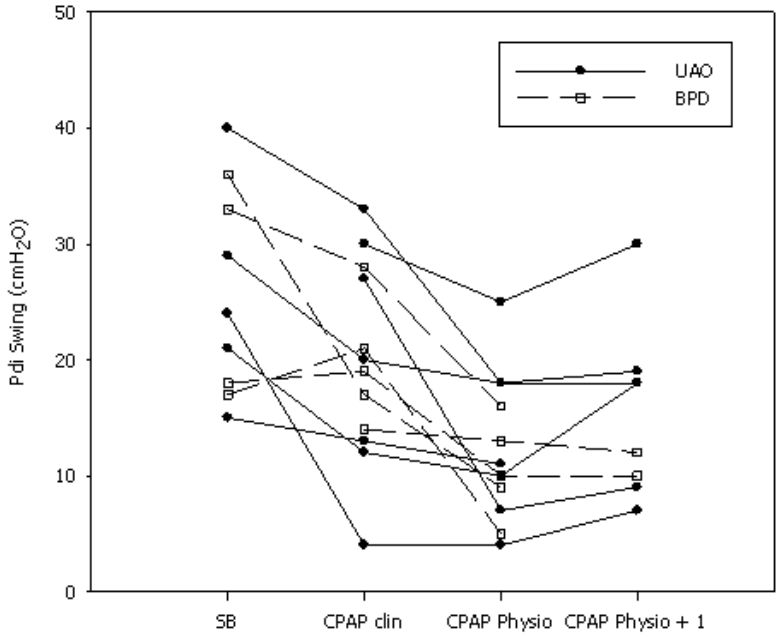
Intensive Care Medicine 2012

Continuous positive airway pressure titration in infants with severe upper airway obstruction or bronchopulmonary dysplasia

Sonia Khirani[1,2], Adriana Ramirez[2,3], Sabrina Aloui[2], Nicolas Leboulanger[4,5,6], Arnaud Picard[5,7] and Brigitte Fauroux[2,5,6*]

CRITICAL CARE



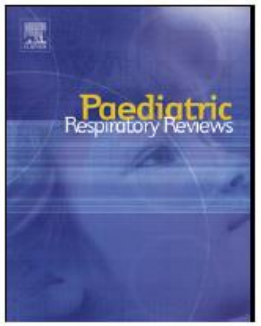


NIV in Pediatrics

- Which patients ?
- When to start ?
- Where to start ?
- Follow up and weaning ?

Oxygen and carbon dioxide monitoring during sleep

Alessandro Amaddeo, Brigitte Fauroux *



No validated (against end-organ morbidity) definition of alveolar hypoventilation in children

Table 2
Definitions of nocturnal hypercapnia or alveolar hypoventilation used in the literature.

Definitions	References
• a percentage of sleep time with a $PtcCO_2 > 50$ mmHg $> 25\%$	• Berry RB, Sleep Med 2012
• a 95 percentile of $PtcCO_2 > 50$ mmHg	• Ward S, Thorax 2005
• a percentage of sleep time with a $PtcCO_2 > 50$ mmHg $> 2\%$	• Paiva R, Intensive Care Med 2009
• a $PtcCO_2 > 55$ mmHg > 10 minutes	• Aboussouah LS, Resp Crit Care Med 2015
• a peak $PtcCO_2 > 49$ mmHg	• Nardi J, Resp Care 2012
• a mean $PtcCO_2 > 50$ mmHg	• Simonds AK, Eur Resp Rev 2013
• a $PtcCO_2 > 10$ mmHg above waking baseline level	• O'Donaghue FJ, ERJ 2003

Abbreviations: $P_{ET}CO_2$: exhaled carbon dioxide; $PtcCO_2$: transcutaneous carbon dioxide.

SPECIAL ARTICLES

JCSM
Journal of Clinical Sleep Medicine
http://dx.doi.org/10.5664/jcsm.2172

OSA

Rules for Scoring Respiratory Events in Sleep: Update of the 2007 AASM Manual for the Scoring of Sleep and Associated Events

Deliberations of the Sleep Apnea Definitions Task Force of the American Academy of Sleep Medicine

study). For adults, sleep hypoventilation is scored when the arterial PCO_2 (or surrogate) is > 55 mm Hg for ≥ 10 minutes or there is an increase in the arterial PCO_2 (or surrogate) ≥ 10 mm Hg (in comparison to an awake supine value) to a value exceeding 50 mm Hg for ≥ 10 minutes. For pediatric patients hypoventilation is scored when the arterial PCO_2 (or surrogate) is > 50 mm Hg for > 25% of total sleep time. In adults Cheyne-Stokes breathing is scored when both of

When to start CPAP ?

- No validated criteria: lack of validated markers of OSA-end-organ morbidity in children
- Recommendations:

TASK FORCE REPORT
ERS STATEMENT

Obstructive sleep disordered breathing in 2- to 18-year-old children: diagnosis and management

Athanasios G. Kaditis[1], Maria Luz Alonso Alvarez[2], An Boudewyns[3], Emmanouel I. Alexopoulos[4], Refika Ersu[5], Koen Joosten[6], Helena Larramona[7], Silvia Miano[8], Indra Narang[9], Ha Trang[10], Marina Tsaoussoglou[1], Nele Vandenbussche[11], Maria Pia Villa[12], Dick Van Waardenburg[13], Silke Weber[14] and Stijn Verhulst[15]

6.7. What are the indications, efficacy and potential complications of CPAP or NPPV in children with obstructive SDB?

Summary

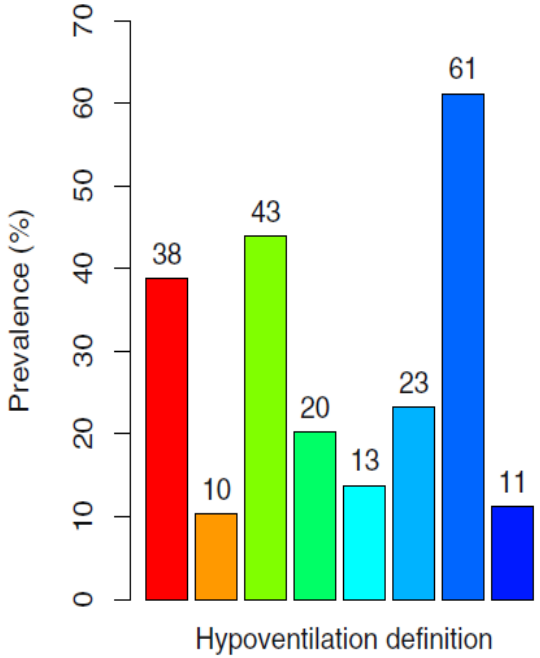
a) Usual indications for CPAP are: residual OSAS after adenotonsillectomy (AHI >5 episodes·h^{-1}) and OSAS related to obesity, craniofacial abnormalities or neuromuscular disorders. If nocturnal hypoventilation occurs (*e.g.* end-tidal carbon dioxide tension (P_{CO_2}) >50 mmHg for >25% of total sleep time or peak end-tidal P_{CO_2} ⩾55 mmHg) NPPV is preferred.

Sleep Breath (2016) 20:575–581
DOI 10.1007/s11325-015-1247-2

SLEEP BREATHING PHYSIOLOGY AND DISORDERS • ORIGINAL ARTICLE

Nocturnal hypoventilation in neuromuscular disease: prevalence according to different definitions issued from the literature

Adam Ogna[1] · Maria-Antonia Quera Salva[2] · Helene Prigent[3] · Ghassane Mroue[2] · Isabelle Vaugier[4] · Djillali Annane[1] · Frederic Lofaso[2,3] · David Orlikowski[1,4]

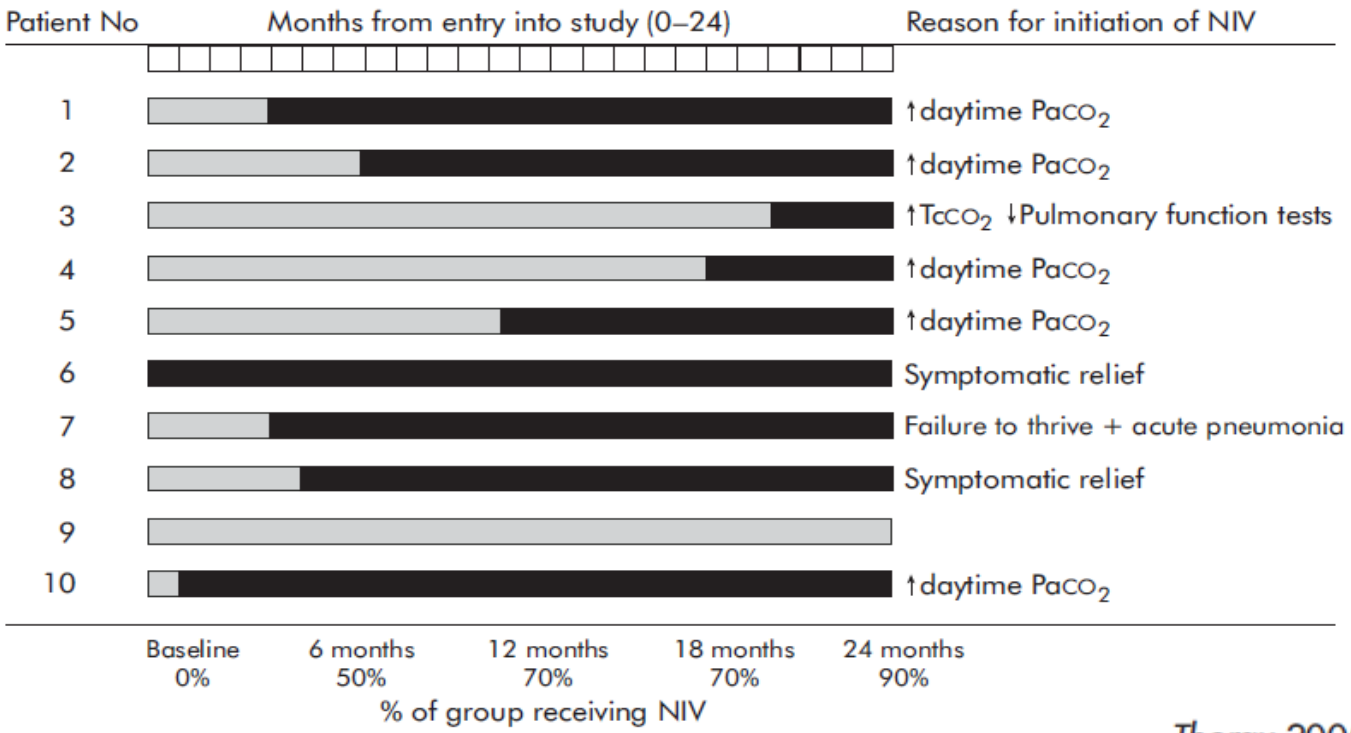


Daytime $PaCO_2$ > 45 mmHg
Daytime base excess > 4 mmmole/L
Nocturnal SpO_2 < 88% for > 5 min
Mean nocturnal SpO_2 < 90% or SpO_2 < 90% for > 10% recording time
$PtcCO_2$ > 55mmHg > 10 min
Increase $PtcCO_2$ > 10 mmHg (awake vs sleep) to a value > 50 mmHg > 10 min
Peak $PtcCO_2$ > 49 mmHg
Mean $PtcCO_2$ > 50 mmHg

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

22 adults and children with normal daytime $PaCO_2$
and a nocturnal peak $PtcCO_2$ > 6.5 kPa (48.75 mmHg)



Thorax 2005;**60**:1019–1024.

When to start NIV ?

- Children with NMD resulting in
  - symptomatic nocturnal hypoventilation or
  - daytime hypercapnia should be supported with NIV

Too late !!!

Hull *et al.* Thorax 2012;67:i1

Nocturnal hypoventilation: predictors and outcomes in childhood progressive neuromuscular disease

Sherri L Katz,[1–3] Isabelle Gaboury,[4] Krista Keilty,[5,6] Brenda Banwell,[6–8] Jiri Vajsar,[7,8] Peter Anderson,[3,9] Andy Ni,[2,3] Ian MacLusky[1,2]

Arch Dis Child 2010;**95**:998–1003.

Table 2 Symptoms at baseline

	Total cohort (n=45)*	NH (n=7)	No NH (n=38)	p Value
Snoring, n (%)				
Always	2 (4.4)	1 (14.3)	1 (2.6)	0.097
Often	2 (4.4)	1 (14.3)	1 (2.6)	
Sometimes	6 (13.3)	2 (28.6)	4 (10.5)	
Occasionally	18 (40.0)	1 (14.3)	17 (44.7)	
Never	17 (37.8)	2 (28.6)	15 (39.5)	
Alert in the morning, n (%)				
Always	16 (35.6)	3 (42.9)	13 (34.2)	0.550
Often	13 (28.9)	1 (14.3)	12 (31.6)	
Sometimes	6 (13.3)	2 (28.6)	4 (10.5)	
Occasionally	5 (11.1)	0 (0)	5 (13.2)	
Never	5 (11.1)	1 (14.3)	4 (10.5)	
Morning headaches, n (%)†				
Never	28 (63.6)	5 (71.4)	23 (62.2)	1.000
1–2 days	13 (28.3)	2 (28.6)	11 (29.7)	
3–4 days	0 (0)	0 (0)	0 (0)	
5–6 days	3 (6.8)	0 (0)	3 (8.1)	
7 days	0 (0)	0 (0)	0 (0)	
Sleepy during the day, n (%)				
Always	0 (0)	0 (0)	0 (0)	1.000
Often	6 (13.3)	1 (14.3)	5 (13.2)	
Sometimes	11 (24.4)	2 (28.6)	9 (23.7)	
Occasionally	16 (35.6)	2 (28.6)	14 (36.8)	
Never	12 (26.7)	2 (28.6)	10 (26.3)	

Table 2 Symptoms at baseline

	Total cohort (n=45)*	NH (n=7)	No NH (n=38)	p Value
Snoring, n (%)				
Number of days with nap, n (%)				
None	34 (75.6)	5 (71.4)	29 (76.3)	0.201
1–2 per week	8 (17.8)	1 (14.3)	7 (18.4)	
3–4 per week	2 (4.4)	0 (0)	2 (5.3)	
5–6 per week	0 (0)	0 (0)	0 (0)	
Every day	1 (2.2)	1 (14.3)	0 (0)	
Number of nocturnal awakenings per night, n (%)†				
Never	20 (45.5)	3 (42.9)	17 (45.9)	0.763
Once	14 (31.8)	3 (42.9)	11 (29.7)	
2–4 times	10 (22.7)	1 (14.3)	9 (24.3)	
5–7 times	0 (0)	0 (0)	0 (0)	
>8 times	0 (0)	0 (0)	0 (0)	
Frequency of care giver attending to child at night, n (%)†				
Never	32 (72.7)	4 (57.1)	28 (75.7)	0.451
1–2 times	11 (25.0)	3 (42.9)	8 (21.6)	
3–4 times	1 (2.3)	0 (0)	1 (2.7)	

Sleep-related hypoventilation was defined as:

- >10 mmHg increase in $PetCO_2$ and/or
- a fall ≥ 5% in SpO_2 for > 10 min

Fleur M. Paulides
Frans B. Plötz
Laura P. Verweij-van den Oudenrijn
Josephus P. J. van Gestel
Mike J. Kampelmacher

Thirty years of home mechanical ventilation in children: escalating need for pediatric intensive care beds

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)

*Data only available for 141 patients.
OSA, obstructive sleep apnoea; CPAP, continuous positive airway pressure; FVC, forced vital capacity

Intensive Care Med (2012) 38:847–852

Long Term Continuous Positive Airway Pressure (CPAP) and Noninvasive Ventilation (NIV) in Children: Initiation Criteria in Real Life

A. Amaddeo, MD,[1,2,3] J. Moreau, MD,[1,4] A. Frapin, MSN,[1] S. Khirani, PhD,[1,5] O. Felix, MD,[1,6] M. Fernandez-Bolanos, MSc,[1] A. Ramirez, MSc,[1,7] and B. Fauroux, MD, PhD[1,2,3]*

Acute
Impossibility to wean from NIV in the ICU
N=15

Subacute
Abnormal nocturnal gas exchange
N=18

Chronic
Abnormal P(S)G
N=43

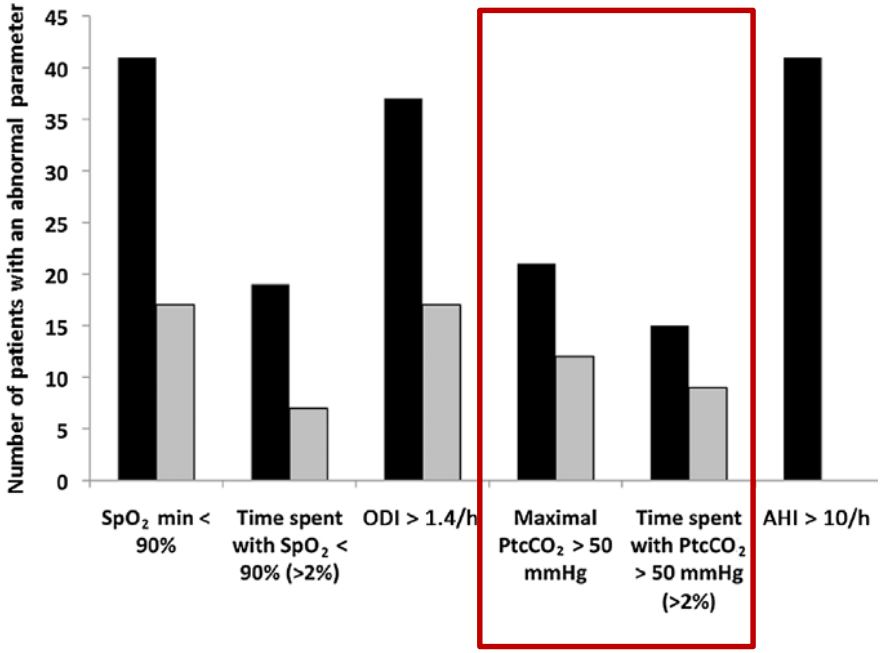
TABLE 2—Respiratory Variables Used for Continuous Positive Pressure or Noninvasive Ventilation Initiation

1. Minimum SpO_2 <90%
2. Maximal $PtcCO_2$ >50 mmHg
3. Time spent with a SpO_2 <90% ≥2% of recording time
4. Time spent with a $PtcCO_2$ >50 mmHg ≥2% of recording time
5. Oxygen desaturation index >1.4 events/hr
6. AHI >10 events/hr

Pediatric Pulmonology 51:968–974 (2016)

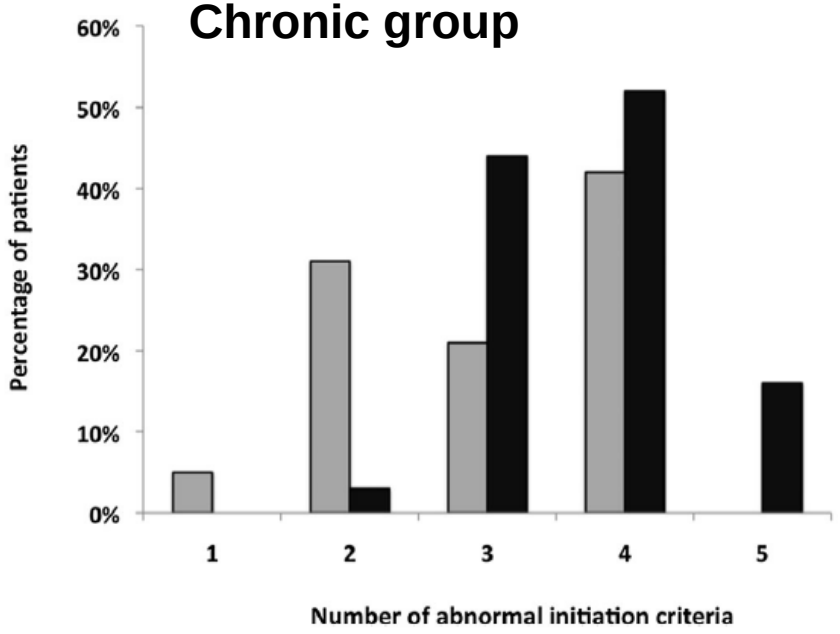
Long Term Continuous Positive Airway Pressure (CPAP) and Noninvasive Ventilation (NIV) in Children: Initiation Criteria in Real Life

A. Amaddeo, MD,[1,2,3] J. Moreau, MD,[1,4] A. Frapin, MSN,[1] S. Khirani, PhD,[1,5] O. Felix, MD,[1,6] M. Fernandez-Bolanos, MSc,[1] A. Ramirez, MSc,[1,7] and B. Fauroux, MD, PhD[1,2,3]*



Subacute group

Chronic group



Pediatric Pulmonology 51:968–974 (2016)

NIV initiation: scenarios

- Castro-Codesal (Alberta)
  - 73%: electively (on PSG)
  - 17%: acute illness
  - 2%: FVC < 30%, transition from IV, palliative care

Table 5. Longitudinal trends in the technology for children using long-term non-invasive ventilation.

	Jan 2005-Apr 2008 (n = 127)	May 2008-Aug 2011 (n = 262)	Sept 2011-Dec 2014 (n = 233)	*P* Value
Trigger for NIV [a]; n, %				
Electively with PSG	90, 71 (95%CI 62–78)	193, 74 (95%CI 68–79)	170, 73 (95%CI 67–78)	0.85
Electively without PSG	19, 15 (95%CI 10–22)	22, 8 (95%CI 6–12)	18, 8 (95%CI 5–12)	0.05
Acute illness	16, 13 (95%CI 8–19)	43, 17 (95%CI 12–21)	40, 17 (95%CI 13–23)	0.48
Other [b]	2, 1 (95%CI 0.4–6)	2, <1 (95%CI 0.2–3)	4, 2 (95%CI 0.6–4)	0.62

- Chatwin (Royal Brompton hospital, UK)
  - 15% started in the PICU

NIV in Pediatrics

- Which patients ?
- When to start ?
- Where to start ?
- Follow up and weaning ?

NIV initiation: hospital or home ?

- Castro-Codesal (Alberta)

	Jan 2005-Apr 2008 (n = 127)	May 2008-Aug 2011 (n = 262)	Sept 2011-Dec 2014 (n = 233)	*P* Value
Location to start NIV [a]; n, %				
Home settings	107, 84 (95% CI 77–90)	214, 82 (95%CI 77–86)	185, 79 (95%CI 74–84)	0.49
PICU	6, 5 (95%CI 2–10)	26, 10 (95%CI 7–14)	26, 12 (95%CI 0.8–16)	0.1
Ward	14, 11 (95%CI 6–18)	21, 8 (95%CI 5–12)	21, 9 (95%CI 6–13)	0.63

- Chatwin (Royal Brompton hospital, UK)
  - 76% started as in-patients
- Necker
  - Majority started as in-patients
- Conclusion
  - No comparative study
  - Depends on patient's age, underlying disease, medical condition, family environment, local facilities

NIV in Pediatrics

- Which patients ?
- When to start ?
- Where to start ?
- Follow up and weaning ?

Long Term Non-Invasive Ventilation in Children: Impact on Survival and Transition to Adult Care

Michelle Chatwin[1,3]*, Hui-Leng Tan[2,3], Andrew Bush[2,3], Mark Rosenthal[2,3], Anita Kay Simonds[1,3]

Table 2. Patients under the age of 17 who were commenced and ceased NIV and CPAP by group diagnosis.

Diagnosis Group	Total patients% (n)	Total patients onNIV% (n)	Total patientson CPAP% (n)	Total patients alive and discontinued NIV% (n)	Total patients alive and discontinued CPAP% (n)	Total patients who died using NIV% (n)	Total patientswho died using CPAP % (n)
Chest wall disease	4 (16)	100 (16)	0 (0)	13 (2)	0 (0)	6 (1)	0 (0)
Obesity	3 (12)	75 (9)	25 (3)	8 (1)	16 (2)	0 (0)	0 (0)
Chronic lung disease	5 (24)	87 (21)	13 (3)	25 (6)	0 (0)	21 (5)	4 (1)
Central sleep apnoea	3 (12)	100 (12)	0 (0)	0 (0)	0 (0)	0 (0)	0 (0)
Cardiac surgery	2 (11)	36 (4)	64 (7)	0 (0)	36 (4)	0 (0)	36 (4)
Congenital syndrome	14 (61)	56 (34)	44 (27)	7 (4)	5 (3)	10 (6)	5 (3)
Upper airway abnormality	9 (39)	62 (24)	38 (15)	10 (4)	15 (6)	5 (2)	2.5 (1)
Other	4 (20)	95 (19)	5 (1)	25 (5)	0 (0)	50 (10)	0 (0)
Neuromuscular disease	56 (254)	96.6 (253)	0.4 (1)	2 (4)	0 (1)	30 (76)	0 (0)

Weaning from long term continuous positive airway pressure or noninvasive ventilation in children

Meriem Mastouri MD[1,2] | Alessandro Amaddeo MD[1,3,4] | Lucie Griffon MD[1] | Annick Frapin MSN[1] | Samira Touil BSc[1] | Adriana Ramirez MSc[1,5] | Sonia Khirani PhD[1,5,6] | Brigitte Fauroux MD, PhD[1,3,4]

TABLE 1 Criteria that may allow weaning from continuous positive airway pressure (CPAP) or noninvasive ventilation (NIV)

Major criteria
1) disappearance of nocturnal and daytime symptoms of sleep-disordered breathing after several nights sleeping without CPAP/NIV, such as snoring, sweating, arousals, labored breathing, change in behavior or attention,
2) percentage of recording time spent with a SpO_2 ≤90% <2%,
3) percentage of recording time spent with a $PtcCO_2$ ≥50 mmHg <2%,
4) obstructive apnea-hypopnea index <10 events/h on a poly(somno)graphy
Minor criteria
1) minimal SpO_2 >90%
2) maximal $PtcCO_2$ <50 mmHg
3) oxygen desaturation index ≤1.4 events/h.

Male/Female	38/20	
Age at CPAP/NIV initiation (years), median (range)	1.4 (0.01-16.2)	
Ventilatory mode, *n* (%)		
CPAP	50 (86%)	
NIV	8 (14%)	
Duration of CPAP/NIV (year), median (range)	1.12 (0.16-8.85)	
CPAP	0.98 (0.16-7.49)	
NIV	3.96 (0.27-8.85)	
Primary diagnosis, *n* (%)	Laryngeal obstruction[a]	9 (16%)
	Pierre Robin syndrome	6 (10%)
	Prader Willi syndrome	6 (10%)
	Treacher Collins syndrome	6 (10%)
	Bronchopulmonary dysplasia	4 (7%)
	Achondroplasia	3 (5%)
	Idiopathic OSA	2 (3%)
	Craniofaciostenosis (Crouzon, Apert)	2 (3%)
	Pycnodysostosis	2 (3%)
	Mucopolysaccharidosis	2 (3%)
	Polymalformative syndrome	2 (3%)
	Mandibular hypoplasia	2 (3%)
	Lung sequelae of viral infection/ARDS	2 (3%)
	Other[b]	10 (21%)
Reason of CPAP/NIV weaning, *n* (%)	Spontaneous improvement:	33 (57%)
	Switch to oxygen therapy:	2 (3%)
	After surgery:	
	ENT surgery	14 (24%)
	Maxillofacial surgery	6 (11%)
	Neurosurgery	1 (2%)
	ENT and neurosurgery	2 (3%)

Pediatric Pulmonology. 2017;1–6.

NIV in Pediatrics

- Which patients
  - Those with nocturnal « alveolar hypoventilation » (persisting after optimal CPAP in case of OSA)
  - > Restrictive/NMD, central hypoventilation
- When to start
  - Nocturnal « alveolar hypoventilation »
  - Importance of screening before an acute exacerbation
- Where to start
  - Hospital or home
- Follow up and weaning
  - Follow up in pediatric expert multidiscilonary center
  - A minority can be weaned from NIV