



Brigitte Fauroux

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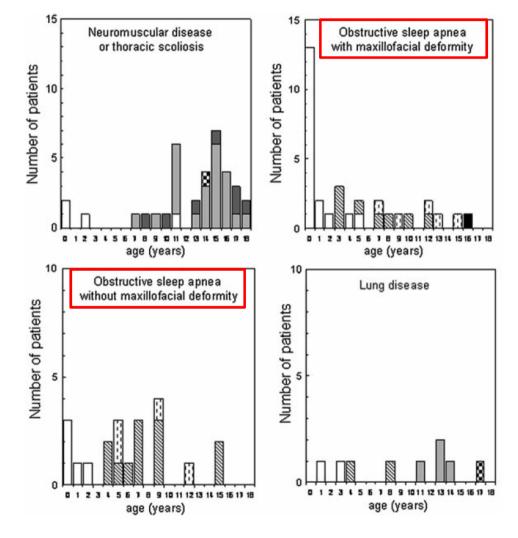






- For which patients ?
- When to start ?
- Which CPAP mode & setting ?
- Which equipment ?
- How to monitor ?
- When to stop ?
- Conclusion

Patients started on home CPAP



Ramirez et al. Intensive Care Med 2012;38:655

Polygraphic respiratory events during sleep in children treated with home continuous positive airway pressure: description and clinical consequences

Johan Moreau ^{a,d,e}, Adriana Ramirez ^{a,f}, Sonia Khirani ^{a,g}, Brigitte Fauroux ^{a,h,i,*} Alessandro Amaddeo ^{a,b}, Valeria Caldarelli ^c, Marta Fernandez-Bolanos ^a,

	ŝ	ŝ	ŝ	ŝ	2	2	1		1	1	1	1	1	1	1	1	1
6/20 7.8 ± 6.2 10.6 ± 14.4	- Down syndrome	- Treacher Collins syndrome	 Polymalformative syndrome 	- Idiopathic OSAS	- Achondroplasia	 CATCH-22 syndrome 	 Neurofibromatosis type 1 with 	subglottic neurofibroma	- Bronchopulmonary dysplasia	- Turner syndrome	 Menkes syndrome 	- Cherubism	- Beckwith-Wiedemann syndrome	- Pycnodysostosis	- Niemann-Pick disease type A	 Post-intubation laryngeal paralysis 	- Prader-Willi syndrome
Female to male ratio (F/M) Age, years (mean ± SD) Time of follow-up, months (mean ± SD)	Predisposing conditions																

Sleep Medicine 16 (2015) 107-112

Rev Port Pneumol. 2014;20(3):146-151



ORIGINAL ARTICLE

Non-invasive ventilation in complex obstructive sleep apnea – A 15-year experience of a pediatric tertiary center



I.C. Girbal*, C. Gonçalves, T. Nunes, R. Ferreira, L. Pereira, A. Saianda, T. Bandeira

Non-invasive ventilation in complex obstructive sleep apnea

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Table 1 Distribution of patients according to primary diagnosis, presence of associated hypoventilation and age at NIV start.	o primary diagnosis, pi	esence of associated hypoven	tilation and age at NIV start.
Nosologic group	Patients (N = 68)	OSA plus hypoventilation syndrome	Age at NIV start in months
	n (%) ^b	n (%) h	Median (IQR)
Congenital malformations/genetic disorders	34 (50)	7 (10)	42.5 (5-144)
Prader-Willi syndrome	9	2	176 (158-187)
Pierre-Robin syndrome	5	0	1 (0-2)
Trisomy 21	5	2	120 (46-180)
Craniofacial malformation ^a	10	0	40 (7-45)
Airway malacia	5	0	13 (2-15)
Other	3	3	60 (40-96)
Cerebral palsy	9 (13)	2 (3)	168 (89-173)
Central nervous system tumor	8 (12)	1 (1.5)	171 (94–180)
Inborn errors of metabolism	6 (6)	2 (3)	59 (20-135)
Mucopolysaccharidosis	5	2	59 (46-156)
Gaucher disease	1	0	2
Adenoid/tonsil hypertrophy	3 (4)	0	15 (12-31)
Obesity	3 (4)	0	166 (154-194)
Others	5 (8)	0	106 (85-110)

TASK FORCE REPORT ERS STATEMENT

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



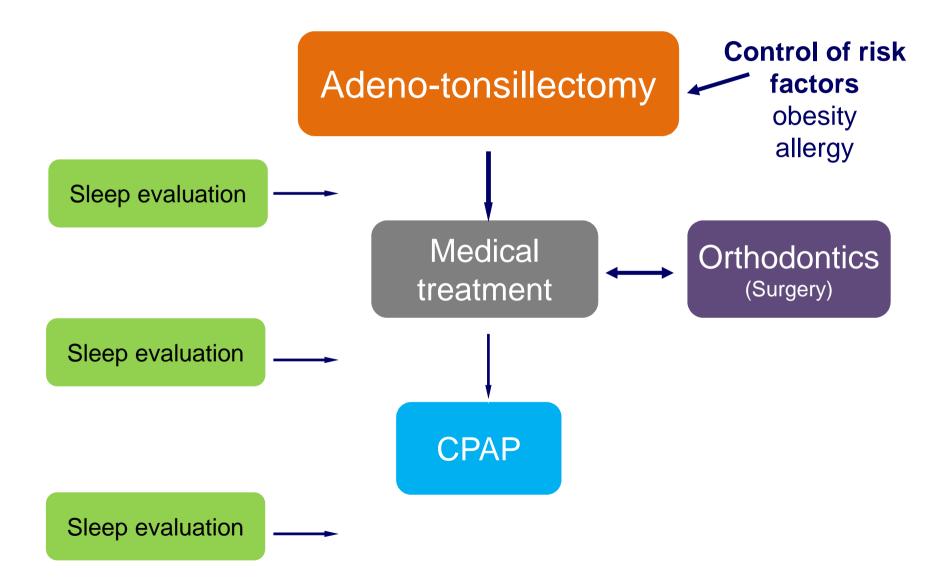
Athanasios G. Kaditis¹, Maria Luz Alonso Alvarez², An Boudewyns³, Emmanouel I. Alexopoulos⁴, Refika Ersu⁵, Koen Joosten⁶, Helena Larramona⁷, Silvia Miano⁸, Indra Narang⁹, Ha Trang¹⁰, Marina Tsaoussoglou¹, Nele Vandenbussche¹¹, Maria Pia Villa¹², Dick Van Waardenburg¹³, Silke Weber¹⁴ and Stijn Verhulst¹⁵ 5.3. Are there conditions predisposing to upper airway obstruction which make treatment of obstructive SDB a priority? Summary

- a) Major craniofacial abnormalities
- b) Neuromuscular disorders
- c) Achondroplasia
- d) Chiari malformation
- e) Down syndrome
- f) Mucopolysaccharidoses
- g) Prader-Willi syndrome



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When to start CPAP ?



When to start CPAP ?

- No validated criteria: lack of validated markers of OSA-end-organ morbidity in children
 TASK FORCE REPORT FRS STATEMENT
- Recommendations:



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

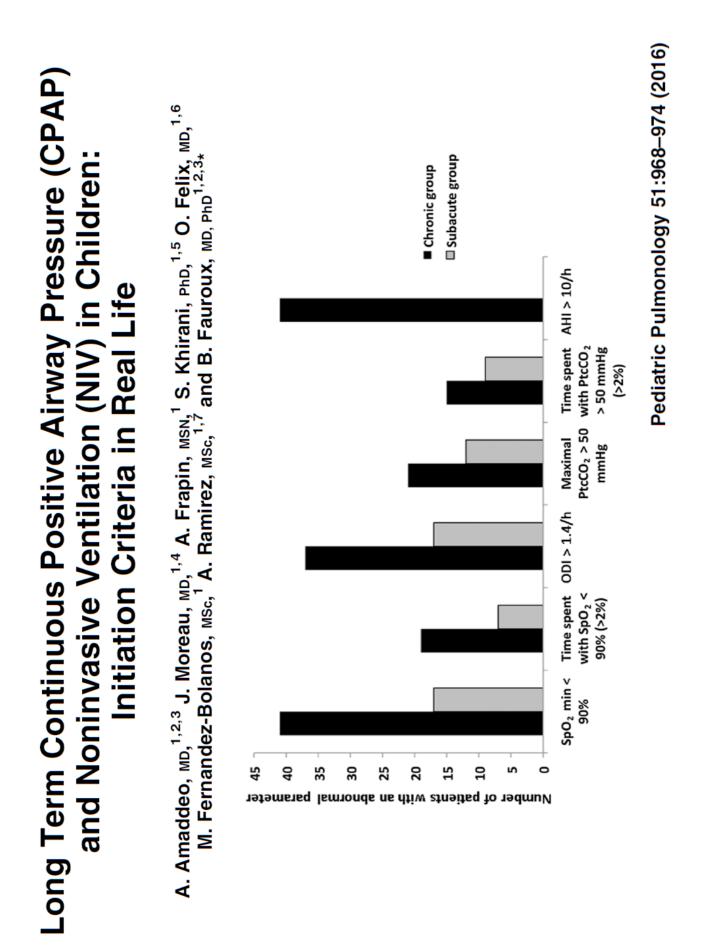
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6.7. What are the indications, efficacy and potential complications of CPAP or NPPV in children with obstructive SDB?

Summary

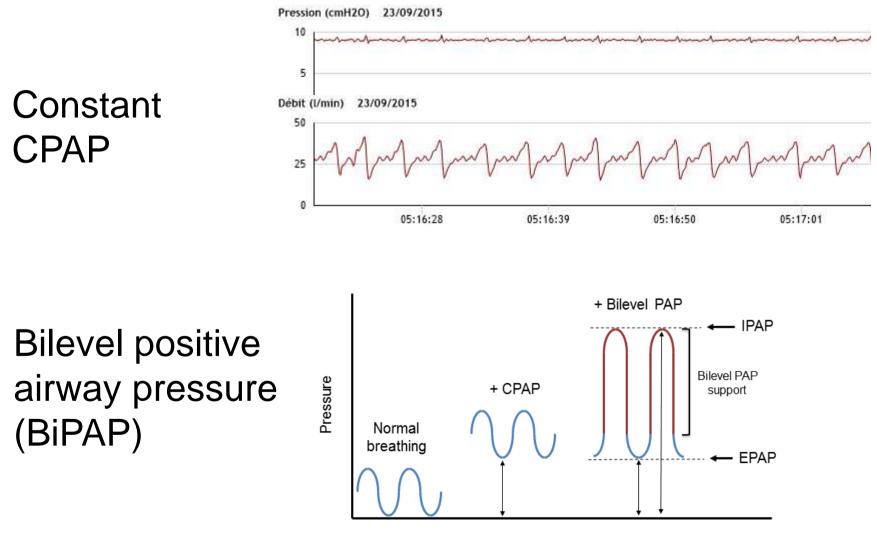
a) Usual indications for CPAP are: residual OSAS after adenotonsillectomy (AHI >5 episodes·h⁻¹) and OSAS related to obesity, craniofacial abnormalities or neuromuscular disorders. If nocturnal hypoventilation occurs (*e.g.* end-tidal carbon dioxide tension (PCO_2) >50 mmHg for >25% of total sleep time or peak end-tidal $PCO_2 \ge 55$ mmHg) NPPV is preferred.

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} *	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 $SpO_2 < 90\% \ge 2\%$ of recording time	4. Time spent with a PtcCO ₂ >50 mmHg $\geq 2\%$ of recording time	5. Oxygen desaturation index >1.4 events/hr	6. AHI > 10 events/hr	

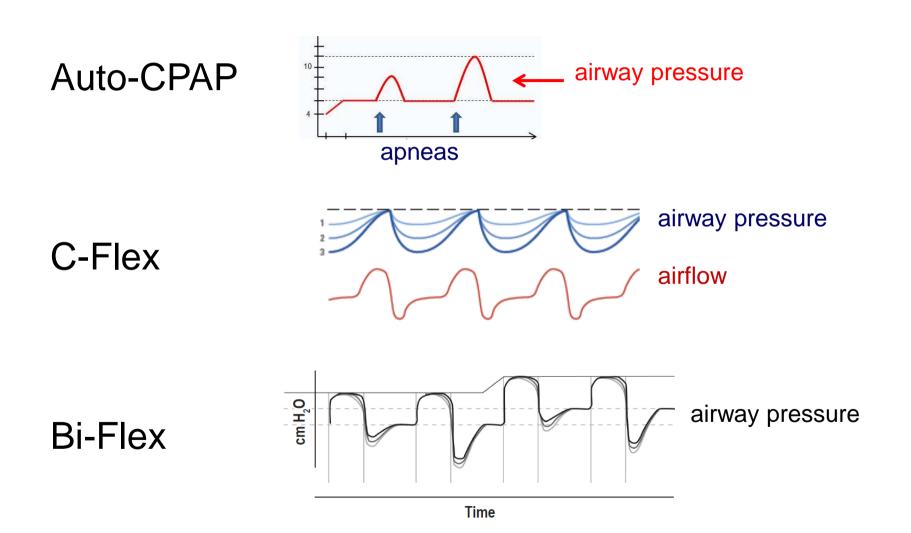


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CPAP modes - 1



CPAP modes - 2



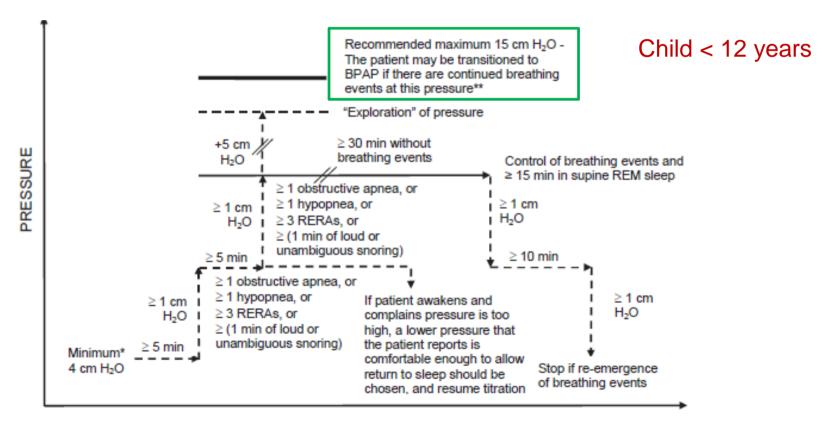


SPECIAL ARTICLE

Clinical Guidelines for the Manual Titration of Positive Airway Pressure in Patients with Obstructive Sleep Apnea

Positive Airway Pressure Titration Task Force of the American Academy of Sleep Medicine

Task Force Members: Clete A. Kushida, M.D., Ph.D., RPSGT (Chair)¹; Alejandro Chediak, M.D. (Vice-Chair)²; Richard B. Berry, M.D.³; Lee K. Brown, M.D.⁴; David Gozal, M.D.⁵; Conrad Iber, M.D.⁶; Sairam Parthasarathy, M.D.⁷; Stuart F. Quan, M.D.⁸; James A. Rowley, M.D.⁹



TIME

Kushida et al. J Clin Sleep Med;2008:4:157

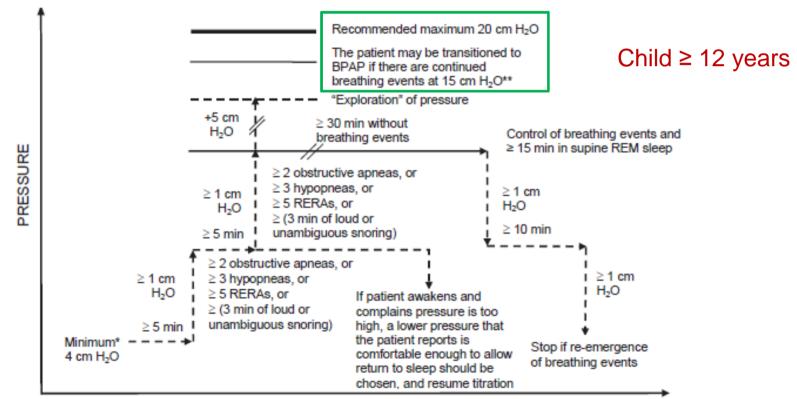


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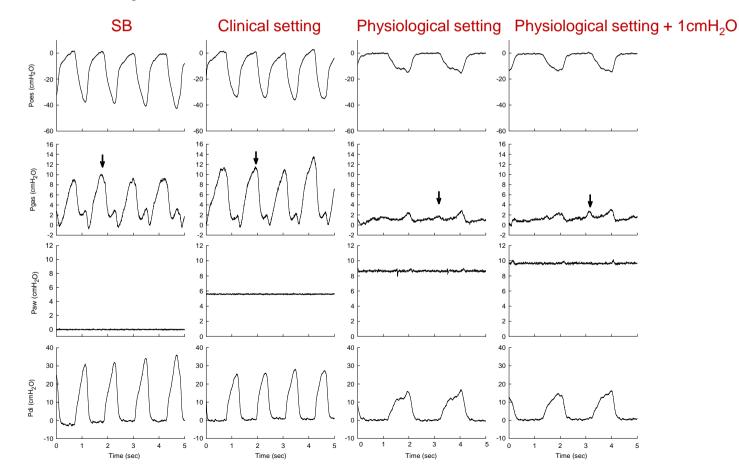


RESEARCH

Open Access

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

Sonia Khirani^{1,2}, Adriana Ramirez^{2,3}, Sabrina Aloui², Nicolas Leboulanger^{4,5,6}, Arnaud Picard^{5,7} and Brigitte Fauroux^{2,5,6*}



Khirani *et al. Critical Care* 2013, **17**:R167 http://ccforum.com/content/17/4/R167

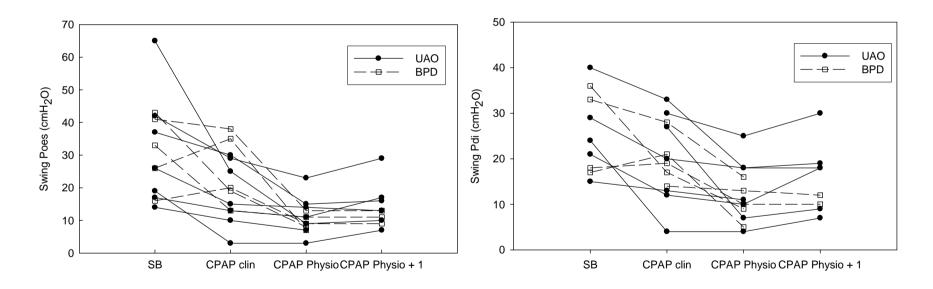


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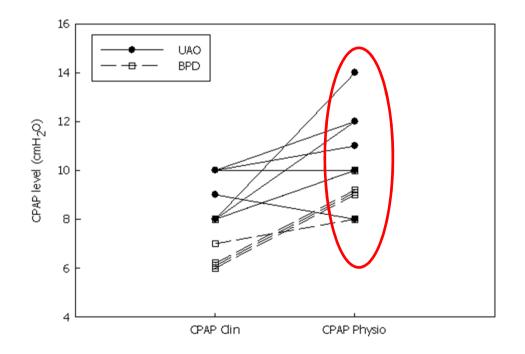


RESEARCH

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

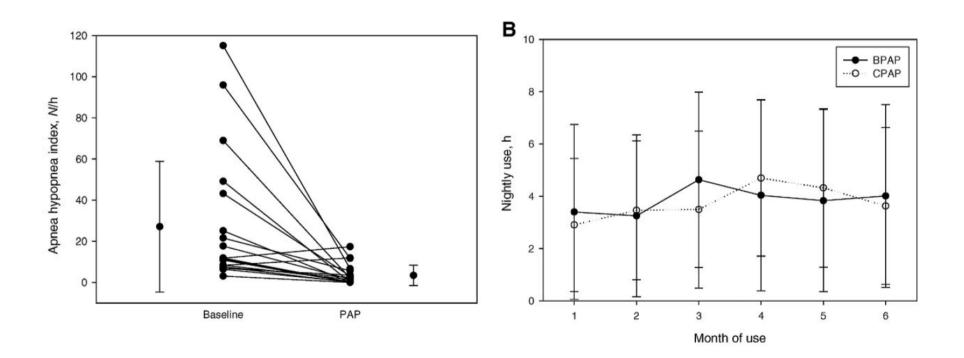
Sonia Khirani^{1,2}, Adriana Ramirez^{2,3}, Sabrina Aloui², Nicolas Leboulanger^{4,5,6}, Arnaud Picard^{5,7} and Brigitte Fauroux^{2,5,6*}



Khirani et al. Crit Care 2013;17:R167

No difference between CPAP and BiPAP

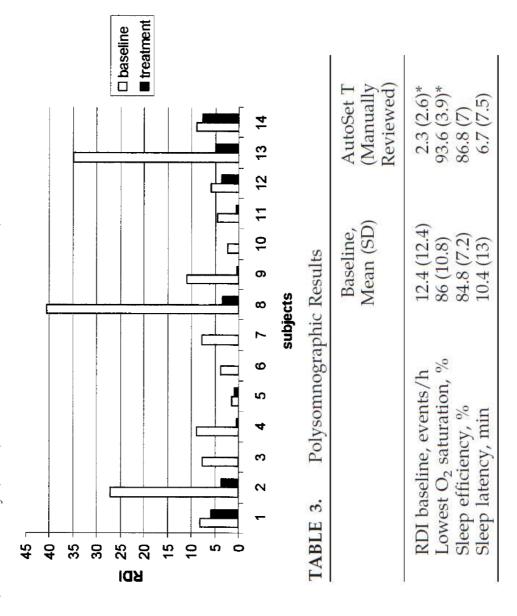
29 children with OSAS: CPAP or Bilevel PAP 1/3 not compliant at 6 months, mean use/night: 5.3 ± 2.5 hours



Marcus et al. Pediatrics 2006;117:e442



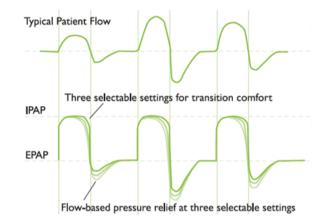
Luciana Palombini, MD; Rafael Pelayo, MD; and Christian Guilleminault, MD



PEDIATRICS Vol. 113 No. 5 May 2004

No difference between CPAP and A-Flex

Randomized double blind trial: CPAP vs Bi-Flex PSG before and after 3 months Objective compliance at 1 and 3 months



56 children and adolescents	СРАР	A-Flex	р
Mean use (nights/month) at M1	24 ± 6	22 ± 9	NS
Mean use (min/night) at M1	201 ± 135	185 ± 165	NS
AHI/h at baseline	22 ± 21	24 ± 6	NS
IAH/h at M1	2 ± 3	2 ± 2	NS
Epworth Sleepiness Scale at baseline	8 ± 5	10 ± 6	NS
Epworth Sleepiness Scale at M1	6 ± 3	5 ± 5	NS

Marcus et al. J Clin Sleep Med 2012;8:37

In practice

- No CPAP mode has proven to be superior to constant CPAP
- Titration
 - in-hospital overnight titration = gold standard
 - alternatively
 - start with auto-CPAP
 - switch to constant CPAP after one week (in-built software)
 - infant: set (progressively) to the highest tolerated level

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Devices able to deliver CPAP













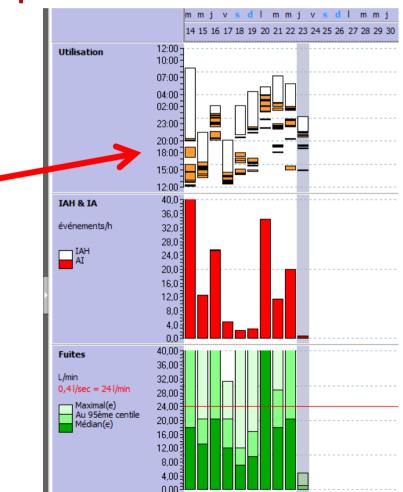


Devices able to deliver CPAP

Devices	CPAP	Auto CPAP	Bilevel PAP	Adjustable trigger	Volume guarantee	Minimal weight	Minimal flow detection
S10 VPAP ST	yes	no	yes	yes	no	13kg	50ml
S10 Autoset	yes	yes	no	no	no	30kg	100ml
BIPAP A30	yes	no	yes	no	200ml	10kg mode ST 20kg AVAPS	Autotrack
BIPAP A40	yes	no	yes	yes	200ml	10kg mode ST 20kg AVAPS	Autotrack or airflow
PR1 REMstar	yes	yes	no	no	no	30kg	Autotrack
PR1 BIPAP ST	yes	no	yes	no	no	18kg	Autotrack
PR1 BIPAP ST AVAPS	yes	no	yes	no	200ml	18kg mode ST 20kg AVAPS	Autotrack
Trilogy	yes	no	yes	yes	50ml	5kg	Autotrack or airflow

Choice of the CPAP device

- Characteristics of the patient
 - battery ? alarms ? (humidification)
 - necessity to use the in-built software ?
- In practice
 - simplicity
 - ergonomy
 - humification



Interfaces for children

Long-term non-invasive ventilation in children

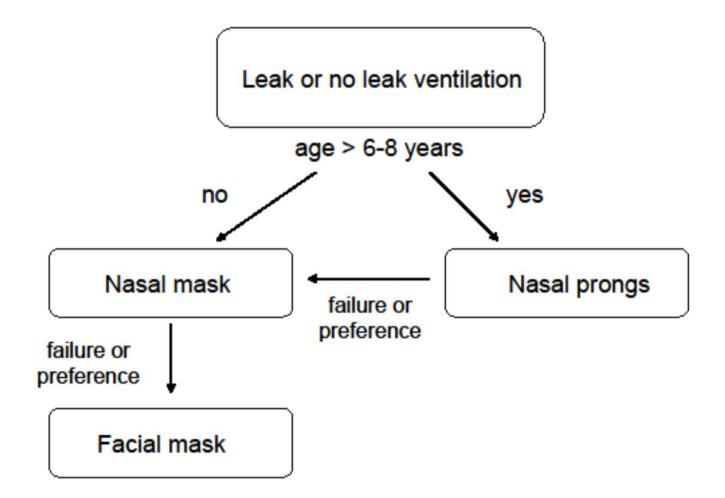


Alessandro Amaddeo, Annick Frapin, Brigitte Fauroux

	Advantages	Disadvantages	Side-effects
Nasal mask	Small internal volume; large choice of different industrial models	Not usable in case of mouth leaks	Pressure sores, eye irritation if leaks, facial deformity
Nasobuccal mask	Prevents mouth leaks	Large volume; risk of inhalation of gastric content in case of gastro-oesophageal reflux; impairs communication and vocalisation; increased aerophagia	Pressure sores, eye irritation if leaks, facial deformity
Total face mask	Prevents mouth leaks	Larger volume than nasobuccal mask; risk of inhalation of gastric content in case of gastro-oesophageal reflux; impairs communication and vocalisation; increased aerophagia	Pressure sores, facial deformity
Nasal pillows	Small and light; no pressure sores	Not usable in case of mouth leaks	Nasal irritation
Mouthpiece	Small and light; no pressure sores; can be used intermittantly	Not useable during sleep	None

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

Choice of the interface



Nasal interfaces for infants













Interfaces for older children



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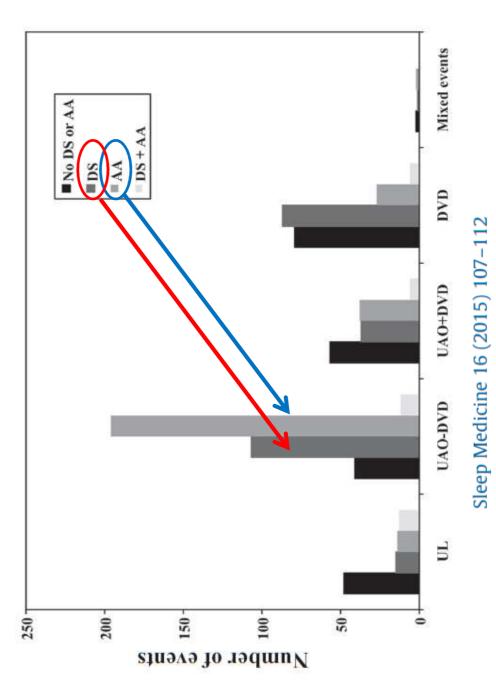
Polygraphic respiratory events during sleep in children treated with home continuous positive airway pressure: description and clinical consequences	r events durin ive airway pr	ıg sleep in cl essure: desc	nildren treate ription and c		sleepmedicine
Alessandro Amaddeo ^{a,b} , Valeria Caldarelli ^c , Marta Fernandez-Bolanos ^a , Johan Moreau ^{a,d,e} , Adriana Ramirez ^{a,f} , Sonia Khirani ^{a,g} , Brigitte Fauroux ^{a,h,i,*}	ria Caldarelli ^c , M. mirez ^{af} , Sonia Kl	arta Fernandez- nirani ^{ag} , Brigitte	·Bolanos ^a , e Fauroux ^{a,h.i,*}		
Polygraphy du Mean CPAP lev Nocturnal gas Mean SpO ₂ (Minimal SpO Time spent v Oxygen Dess Mean PtcCO; Maximal Ptc Time spent v Objective CPAI Average use Average use	Polygraphy duration (h:min) Mean CPAP level (cmH ₂ O) Mocturnal gas exchange Mean SpO ₂ (%) Minimal PtcCO ₂ (mmHg) Maximal Pt	(%) (number/h) nmHg (%) er the last month (n in) hts/month)		7:10 \pm 1:30 7.7 \pm 1.5 97 \pm 1 89 \pm 6 0.2 \pm 0.5 3.8 \pm 5.2 40 \pm 4 40 \pm 4 46 \pm 7 1.7 \pm 6.0 7:40 \pm 2:10 28 \pm 2 28 \pm 2	
Occurrence of SomnoNIV respiratory events during CPAP (<i>n</i> = 29). Unintentional k	ing CPAP (n = 29). Unintentional leaks	Partial or total UAO without decrease in ventilatory drive	Partial or total UAO with decrease in ventilatory drive	Decrease in ventilatory drive	/ Mixed events
Number of polygraphies with the event, <i>n</i> (percentage) (percentage) Event index/h, median (range) Percentage of time spent with each event, median (range)	12 (41%) 0.0 (0.0–3.1) 0.0 (0.0–42.4)	19 (65%) 0.4 (0.0–7.9) 0.7 (0.0–13.7)	13 (45%) 0.0 (0.0–4.8) 0.0 (0.0–7.4)	12 (41%) 0.0 (0.0–25.2) 0.0 (0.0–5.3)	3 (10%) 0.0 (0.0–2.0) 0.0 (0.0–4.0)

Sleep Medicine 16 (2015) 107-112

Polygraphic respiratory events during sleep in children treated with home continuous positive airway pressure: description and clinical consequences

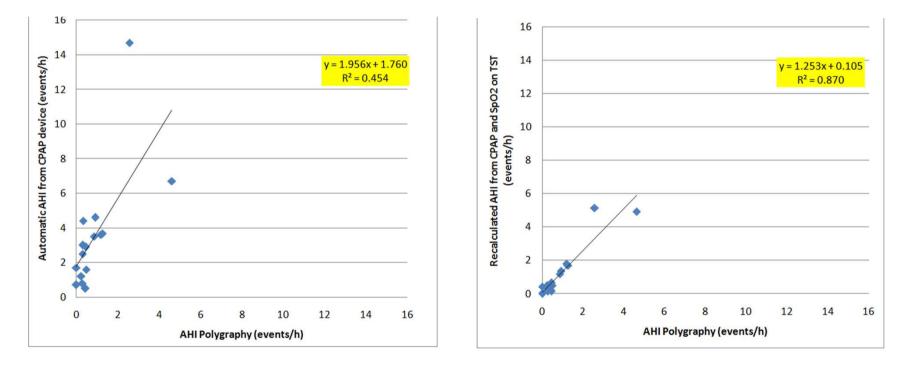
Johan Moreau ^{a,d,e}, Adriana Ramirez ^{a,f}, Sonia Khirani ^{a,g}, Brigitte Fauroux ^{a,h,i,*} Alessandro Amaddeo ^{a,b}, Valeria Caldarelli ^c, Marta Fernandez-Bolanos ^a,





Value of the analysis of the in-built software

Comparison of the data from the in-built software (Rescan) + SpO₂ and a PG



Automatic analysis

Analysis of the in-built software tracings as a PG

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Weaning of CPAP

- Lack of guidelines
- Determinants
 - underlying disease and natural history (PRS)
 - growth, weight loss
 - interventions: orthodontics, surgery...
- Long lasting effect of CPAP
 - importance long term follow up with of regular assessments
- Criteria: normal sleep without CPAP

and	
Noninvasive positive-pressure ventilation avoids recannulation and	hildren*
avoids	ny in cl
ventilation	tracheotom
sitive-pressure	weaning from tracheotomy in children*
ive po	s early
Noninvas	facilitates early wea

Brigitte Fauroux, MD, PhD; Nicolas Leboulanger, MD; Gilles Roger, MD; Françoise Denoyelle, MD, PhD; Arnaud Picard, MD, PhD; Erea-Noel Garabedian, MD; Guillaume Aubertin, MD; Annick Clément, MD, PhD

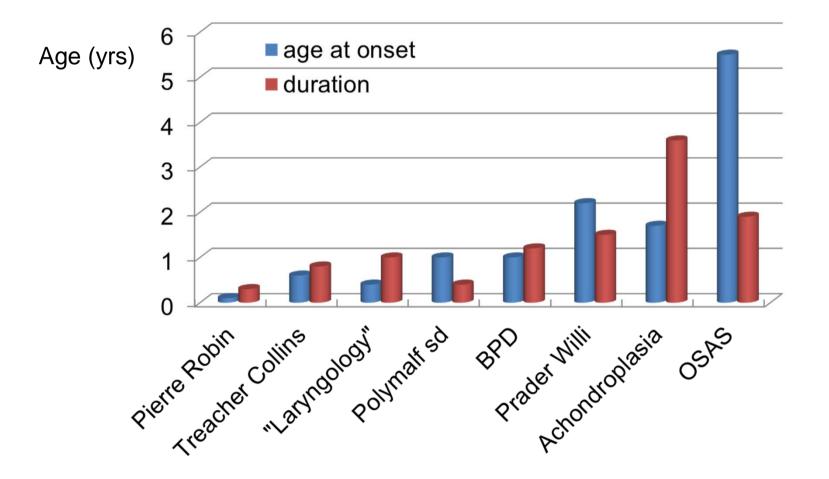
Patient	Gender	Diagnosis	Age at Tracheotomy	Age at Detubation (yrs)	NPPV Delay (mos)	Outcome
Delayed NPPV						
group						
1	female	Treacher-Collins	1 mo	2.5	9	on NPPV since 1 mo
		syndrome			į	
2	female	Vocal cord paralysis +	1 mo	2.5	4	successful NPPV withdrawal at
		tracheomalacia			ļ	age 5, now 7.5 yrs old
3	male	Vocal cord paralysis +	3 mos	11	48	still on NPPV at age 18
		polymalformation				
4	male	Congenital	3 mos	5.7	12	still on NPPV at age 12
		diaphragmatic				
		hypoplasia #			3	
ъ С	female	Cystic lymphangioma	6 mos	2	1	successful NPPV withdrawal at
						age 4, now 11 yrs old
9	male	Vocal cord paralysis +	6 mos	2	1	still on NPPV at age 4
		BPD #				
2	male	Vocal cord paralysis +	6 mos	10	9	still on NPPV at age 11
		multiple congenital				
		anomalies			à	
8	male	Laryngeal cleft	1 yr	3	9	successful NPPV withdrawal at
						age 12, now 13 yrs old
6	female	Vocal cord paralysis + cerebral tumor	6.5 yrs	10.5	6	still on NPPV at age 12

Pediatr Crit Care Med 2010 Vol. 11, No. 1

Weaning of CPAP/NIV 59 patients (25%) / 27 mois

Age at CPAP/NIV initiation (median), yrs	1.4	
CPAP / NIV	51 (86%) / 8 (14%)	
Duration of CPAP / NIV (median), yrs	1 / 4	
Diagnosis	Laryngeal disease Prader Willi sd Bronchopulmonary dysplasia Treacher Collins Pierre Robin sd Polymalformative sd Idiopathic OSA Achondroplasia Crouzon, Apert Pycnodysostosis Mucoplysaccharidosis Goldenhar sd Other	8 6 6 5 5 5 5 3 2 2 2 2 1 6
Reason of withdrawal	Improvement 75%: spontane 2/3, after surgery 1/3 Non compliant, poor tolerance	

Age at onset of CPAP/NIV and duration of treatment



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Conclusion Areas for future research

- Validation of initiation & weaning criteria
- Improvements of the flow detection (inbuilt software) of CPAP devices (infant)
- Nasal prongs and naso-buccal interfaces for infants
- Teletransmission of inbuilt software data + gas exchange (SpO₂ + PtcCO₂)