

**Online Table S3.2:** Initial and follow up settings for CPAP

Author	Country	Journal	Type of study	Number of patients	Ages	CPAP mode	CPAP level	Follow up
Marcus et al. [1]	USA	J Pediatr	Retrospective study (written questionnaire)	94 children with obesity (27%), craniofacial malformation (25%), OSA type I (idiopathic post AT) (17%), Down syndrome (13%)	0-19 yrs	Constant CPAP-titration with PSG	Available for 70 patients Median CPAP =8 (range 4-20), CPAP were independent of age and diagnosis	Every 4 to 12 months Follow up: 22% required modification of CPAP level during follow up
McNamara et al. [2]	Australia	Chest	Prospective study	24 infants on long term CPAP	1-51 weeks old	Constant CPAP - Titration with PSG	Initial setting 3.7 to 6 cmH <sub>2</sub> O Increments of 0.3 until obstruction overcome on PSG 5 infants with upper airway anomalies required up to 10 cmH <sub>2</sub> O	CPAP discontinued in 13 infants ; CPAP level was increased in the 5 other infants (6 drop off due to non-compliance/adherence)
Massa et al. [3]	UK	Arch Dis Child	Retrospective study	42/66 children on long term CPAP (17 (26%) failed trials and alternative treatment)	0-19 yrs	Data on 42 children who ended up successfully	Start at 4 cmH <sub>2</sub> O then increments of 2 cm to overcome OSA and desaturation on PSG Mean CPAP 8.5 ±	Side effect: skin irritation/nasal dryness Up to 3 trials to achieve adherence

						adhering to CPAP	3.2 cmH <sub>2</sub> O (4-16)	
Marcus et al. [4]	USA	Pediatrics	Prospective randomized study: CPAP or BPAP	29 children (13 CPAP 16 BPAP) 19 patients completed the study – of note 19 obese patients	2-16 yrs	CPAP vs BPAP	CPAP started at 3 cmH <sub>2</sub> O then 4 then increments of 2 to overcome OSA on PSG BPAP aim to have 6 differential starting 4/3, 6/3, 8/3, 10/4,12/6,14/8,16/10 CPAP $8 \pm 3$ (4-12) BPAP $11 \pm 4$ (4-16) and $5 \pm 3$ (3-10)	CPAP vs BPAP no effect on drop outs, same mean compliance at 6 m: $5.3 \pm 2.5$ h/night
Tan et al. [5]	Australia	J Pediatr Child Health	Retrospective study over 1 yr	61 sleep studies in 45 children 33% PSG, 33% polygraphy and 33% with autoCPAP	0.4-18.6 yrs (median 8.3 yrs)		64% CPAP - 31% BPAP Changes to improve OSA or ventilation where any persistent apnoea, hypopnoea or hypoventilation	Changes recommended in 66%: 12 CPAP increase, 12 BPAP increase, 1 CPAP decrease, 4 BPAP decrease, 2 CPAP withdrawal
Marcus et al. [6]	USA	J Clin Sleep Med	Prospective double blind randomized study	56 children	2-16 yrs	CPAP vs BiFlex	At 3 m: CPAP $10 \pm 3$ BiFlex $14 \pm 3$ and $8 \pm 2$	At 3 m: same efficacy on AHI and daytime sleepiness and compliance: 24 vs 22 nights/m and 201 vs 185 min/night for CPAP vs

								BiFlex
Khirani et al. [7]	France	Crit Care	Prospective physiological study: oesogastric pressures measures vs clinical parameters, single centre	12 infants, 5 BPD and 7 UAO (3 laryngomalacia, 1 OSA, 1 Down syndrome, 1 Pierre Robin Sequence, 1 Prader Willi syndrome)	2-22 m 3.6-10.3 kg	Constant CPAP, different ventilators	CPAP level set on clinical signs: 8 cmH <sub>2</sub> O CPAP level set on oesogastric pressures: 10 cmH <sub>2</sub> O Physiological data superior to clinical. Patients discharged home with CPAP level determined by physiological data	Follow up program not specified. Improved gas exchange and weight gain. All patients weaned from CPAP (6 m – 3 yo) after improved clinical status
Widger et al. [8]	Australia	Sleep & Breathing	Retrospective study of all patients on respiratory support (CPAP + BPAP) 2007-2012 Single centre	42 children (25 CPAP + 17 BPAP) had 71 PSG	11 ± 6 yrs	CPAP + BPAP	CPAP titration 1-2 cm upwards or downwards based on presence/absence of apnoeas/hypopnoeas on PSG, special protocol for adjustment of BPAP	Annual titration PSGs. Changes recommended in 27/41 studies with CPAP and 11/30 studies with NIV – overall recommended in 53% of studies Full or partial changes implemented in 90% improvement in OSA symptoms on questionnaire in 50% when changes were implemented
Chatwin et al. [9]	UK	PlosOne	Retrospective descriptive study of	449 children started on home NIV,	< 17 yrs 13%<1	CPAP (12%) + BPAP	CPAP settings 8 ± 1.3 cmH <sub>2</sub> O	PSG 3 m after initiation of respiratory support, then 3 m again and if

			outcomes at 1 center 1993-2011	565 with NMD	y, age at initiation 8.7 ±6 y			stable, once a year
Amaddeo et al. [10]	France	Sleep Med	Retrospective study using PGs of consecutive patients between 2011-2014, single centre	29 control PGs in 26 stable children treated with CPAP at home	7.8 ± 6.2 yrs	CPAP in 23 patients and Auto-CPAP in 3	Mean CPAP 7.7 ±1.5cm H <sub>2</sub> O at time of PGs	Median respiratory events index: 1.4/h (range 0-34), > obstructive events often associated with desaturations/arousal; 50% unintentional leaks but with no desaturations. PGs resulted in 7 CPAP changes in settings or interface: 3 increase, 1 decrease, 1 to auto-CPAP, 1 to NIV and 1 interface change
Mihai et al. [11]	Australia	J Clin Sleep Med	Retrospective review on prospective collected data on children initially treated with auto-CPAP before switching to fixed CPAP (2013-2015)	26 children treated with auto-CPAP	11.9 ± 3.4 yrs	Auto-CPAP	Median CPAP level on titration PSG (9 (7-10)) comparable to median 90 <sup>th</sup> percentile CPAP level on auto-CPAP (8.1 (7.1-9.5)) and higher than mean auto-CPAP (6.3 (5.3-7.5))	90 <sup>th</sup> percentile CPAP is useful but does not completely eliminate the need for titration PSG when determining optimal CPAP level. Mean CPAP level downloaded from Auto-CPAP machine can be used to effectively shorten the PSG titration study

Al-Saleh et al. [12]	Canada	J Clin Sleep Med	Retrospective study 2009/2013 Single centre Review of PSGs for technology titration in patients with CPAP, BPAP or IV Major change: changes in mode, pressure/ rate and/or mask Minor change: inspiratory time, rise time, trigger or cycle setting	623 titration PSG in 166 children treated with BPAP and 83 children treated with CPAP and 25 children with IV 50% respiratory disorders, 28% NMD and 22% CNS	10.5 ± 5.1 yrs	CPAP BPAP IV	CPAP titration from 4, increase 1-2, max 15 cmH <sub>2</sub> O Switch to BPAP if CPAP failure BPAP: Start spontaneous/timed mode, titrate from 8/4 cmH <sub>2</sub> O, back-up rate 8 bpm, increase inspiratory / expiratory pressures by 1-2 cmH <sub>2</sub> O, minimum difference 4 cmH <sub>2</sub> O	Major outcome: clinical predictors of changes at follow-up PSG: age at PSG, CNS or NMD diagnosis, BPAP and shorter time between start of therapy and PSG had higher likelihood of a change in settings. 62% major change, 11% minor change, 27% no change, 4% mask change, 3% mode change. First titration study should be done no more than a year after treatment initiation
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Abbreviations: m: months, yrs: years, OSA: obstructive sleep apnea, BPAP: bilevel positive airway pressure, NIV: noninvasive ventilation, IV: invasive ventilation, NMD: neuromuscular disease, PSG: polysomnography, PG: respiratory polygraphy, AHI: apnea-hypopnea index, , CNS: central nervous system.

## References

1. Marcus CL, Ward SL, Mallory GB, *et al.* Use of nasal continuous positive airway pressure as treatment of childhood obstructive sleep apnea. *J Pediatr* 1995; 127: 88-94.

2. McNamara F, Sullivan CE. Obstructive sleep apnea in infants and its management with nasal continuous positive airway pressure. *Chest* 1999; 116: 10-16.
3. Massa F, Gonzalez S, Lavery A, *et al.* The use of nasal continuous positive airway pressure to treat obstructive sleep apnoea. *Arch Dis Child* 2002; 87: 438-443.
4. Marcus CL, Rosen G, Ward SLD, *et al.* Adherence to and effectiveness of positive airway pressure therapy in children with obstructive sleep apnea. *Pediatrics* 2006; 117: e442-e451.
5. Tan E, Nixon GM, Edwards EA. Sleep studies frequently lead to changes in respiratory support in children. *J Paediatr Child Health* 2007; 43: 560-563.
6. Marcus CL, Radcliffe J, Konstantinopoulou S, *et al.* Effects of positive airway pressure therapy on neurobehavioral outcomes in children with obstructive sleep apnea. *Am J Respir Crit Care Med* 2012; 185: 998-1003.
7. Khirani S, Ramirez A, Aloui S, *et al.* Continuous positive airway pressure titration in infants with severe upper airway obstruction or bronchopulmonary dysplasia. *Crit Care* 2013; 17: R167.
8. Widger JA, Davey MJ, Nixon GM. Sleep studies in children on long-term non-invasive respiratory support. *Sleep Breath* 2014; 18: 885-889.
9. Chatwin M, Tan HL, Bush A, *et al.* Long term non-invasive ventilation in children: impact on survival and transition to adult care. *PLoS One* 2015; 10: e0125839.
10. Amaddeo A, Caldarelli V, Fernandez-Bolanos M, *et al.* Polygraphic respiratory events during sleep in children treated with home continuous positive airway pressure: description and clinical consequences. *Sleep Med* 2015; 16: 107-112.
11. Mihai R, Vandeleur M, Pecoraro S, *et al.* Autotitrating CPAP as a tool for CPAP initiation for children. *J Clin Sleep Med* 2017; 13: 713-719.
12. Al-Saleh S, Sayal P, Stephens D, *et al.* Factors associated with changes in invasive and noninvasive positive airway pressure therapy settings during pediatric polysomnograms. *J Clin Sleep Med* 2017; 13: 183-188.