

Online Table S9.3: Special population: CPAP or NIV in children with neurodisability

Author, Year	Journal	Country	Study design	No of patients	Age of patients	Evaluation	Conclusion
Grychtol et al. [1]	Arch Dis Child	UK	Retrospective review of case series (2010-2016)	21 patients with severe CP (20 with GMFCS IV/V)	1.7-16.1 yrs	21 children with CP were moderate – severe SDB were initiated on NIV; Indication for NIV: mod-severe OSA ± hypoventilation, despite upper airway intervention. 11/21 (55%) patients failed to establish on NIV due to mask intolerance ± ventilation pressure at initial trial or poor adherence during follow up. Erratic sleep pattern may have contributed to intolerance of NIV and vice versa. Established NIV users showed good adherence with significant improvement in SDB	Challenging group of patients: success rate less than total NIV patient cohort. Decision to initiate should be based on benefit outweighing risk and burden
Marcus et al. [2]	AJRC CM	USA	Prospective study	52 children with OSA, 10 (19%) had neurodevelopmental disability	12 yr ± 4 yrs	Children treated with CPAP/BIPAP had significant improvement in attention deficit, sleepiness and quality of life. Behavioral factors improved in children with developmental delay.	Heterogeneous group – no subgroup analysis for children with neurodisability. Variable adherence, mean adherence was 3 hours/night.
Hsiao et al. [3]	Res Dev Disabil	New Zealand	Retrospective case control study:	Children with CP (GMCSF	3-18 yr	Treated patients (surgical or CPAP) showed improvement in OSA symptoms (sleep	Treatment of OSA in children with CP leads to significant benefit in aspects of health and QoL.

			comparison between adenotonsillectomy and NIV for OSA with a control group	V) Treatment group (n=10): 7 had AT; 3 on CPAP vs control group (n=9) who had no OSA or treatment		disturbance, daytime functioning, carer's concern)	Limited by small sample, retrospective design and lack of comparison between treatment options
Girbal et al. [4]	Rev Port Pneumol	Portugal	Restrospective cohort, single centre	9 CP (13% of cohort), 6 inborn error of metabolism (9% of cohort)	Age of start of NIV in patients with CP CP 168 m (IQR 89-173)		All patients with sustained NIV had clinical improvement as reported in clinic files. Limitations: complex OSA not defined, patients with CP not analyzed separately

Abbreviations: m: months, yrs: years, CPAP: continuous positive airway pressure, BPAP: bilevel positive airway pressure, NIV: noninvasive ventilation, GMFCS: gross motor function classification system, CP: cerebral palsy, OSA: obstructive sleep apnea, SDB: sleep-disordered breathing, AT: adenotonsillectomy, QoL: quality of life.

References

1. Grychtol R, Chan EY. Use of non-invasive ventilation in cerebral palsy. *Arch Dis Child* 2018; 103: 1170-1177.
2. 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.

3. Hsiao KH, Nixon GM. The effect of treatment of obstructive sleep apnea on quality of life in children with cerebral palsy. *Res Dev Disabil* 2008; 29: 133-140.
4. Girbal IC, Goncalves C, Nunes T, *et al.* Non-invasive ventilation in complex obstructive sleep apnea--a 15-year experience of a pediatric tertiary center. *Rev Port Pneumol* 2014; 20: 146-151.