

Online Table S7: Which options when CPAP or NIV fails?

Author	Country	Journal	Type of study	Number of patients	Ages	Options	Efficacy
Koontz et al. [1]	USA	Sleep	Retrospective	20 children with syndromic OSA not compliant to BPAP	1-15 yrs	Behavioral analysis and then given the options of receiving recommendations or therapy	Improvement of compliance (= hours/night usage) in 75% of patients who received behavioral intervention (recommendation or behavioral therapy); benefit extended beyond the therapy period though follow up period not specified. Not clear what the referral criteria was in terms of non-compliance though at baseline, the usage ranged 0.77-3.3 hours/night - the significance of the duration of use may differ across age groups.
Slifer et al. [2]	USA	Behav Sleep Med	Descriptive case series	4 children prescribed BPAP for complex OSA or hypoventilation but all demonstrated refusal behavior	3-5 yrs	Behavioral therapy	Good use of the device (sleep age-appropriate time) with sustained benefit
Mendoza-Ruiz et al. [3]	France	Sleep Med	Prospective study	6 CPAP non-adherent (defined as <3 hours/night) children and 9 adherent children	2.2-14.1 yrs (non-adherent group); 0.9-8.6 yrs (adherent group)	Use of a table based on token economy, adherence evaluated after 1 mo	Mean adherence in non-adherent children improved from 1 ± 0.33 h/night to 4.31 ± 1.12 h/night at 1 m, but still significantly lower than adherent children
Delord	France	Chest	Retrospective	9 children, in	2-15 yrs	Hypnosis	Acceptance of the NIV in all patients after 1

et al. [4]	e		ctive study	whom initiation of NIV was expected to be difficult or who refused CPAP/NIV by standard procedure (n=2)		Distraction in the 2-yrs old, (In)direct hypnotic suggestions in the other (n=8) older children	session, median of 3 sessions for a > 6h use/night Mean compliance at 6 m: 7.5h/night
Cheng et al. [5]	Australia	ANZ J of Surgery	Retrospective study 2003-2008	20 infants with Pierre Robin Sequence	neonates	CPAP failed (failure to treat respiratory deterioration/distress due to obstruction) in 6 infants: successful management with MDO and glossopexy	MDO with glossopexy were effective for management of multilevel airway obstruction in infants who would otherwise be considered for tracheostomy following failed CPAP
Abel et al. [6]	UK	Arch Dis Child	Retrospective cohort 2000-2010	104 patients with Pierre Robin Sequence	64/104 < 4 weeks of age	Conservative management in 27 patients, NPT in 63 patients, tracheotomy in 14 patients	PSG results improved in all patients with NPT No CPAP trial in any patient.
Kam et al. [7]	Canada	Canadian Respir J	Retrospective cohort	139 patients with Pierre Robin Sequence	9 – 14 m	20 treated with CPAP, 28 NPT, 45 TLA, 5 MDO, 19 tracheotomy	No details on the protocol but 13/60 surgical patients had another intervention prior to surgery TLA was performed earlier than MDO and tracheotomy
Muller-Hagedorn et al.	Germany	Head Face Med	Retrospective study at	68 children with Pierre Robin Sequence	0-12 yrs	Palatal plate	5 patients did not tolerate PP, 2 laryngeal problems, 1 immediate tracheotomy Of the 56 patients who tolerated TPP, 1

[8]			1 center (2003-2009)	treated with PP			needed CPAP and 4 HFNC (with O ₂)
Amaddeo et al. [9]	France	Plastic and Rec Surgery	Retrospective study 1 center (2014-2015)	44 children with Pierre Robin Sequence, 9 received CPAP in NICU	0-2 m	Tracheotomy in 4 out of 9 patients with severe upper airway obstruction and dependent on ventilation in the NICU Failure in CPAP = failure to wean non-invasive CPAP to usage during sleep time only in 1-2 weeks	No objective evaluation of tracheotomy efficacy (not clear if (1) polygraphy post tracheostomy insertion was performed to assess residual obstructive SDB (2) if any patient needed pressure support with tracheostomy in situ (3) no follow up on tracheotomy patients
Overbergh et al. [10]	Belgium	Sleep Med	Case series	9 children with complex OSA and CPAP intolerance	7 m - 15 yrs	Optiflow nasal cannula adapted to a life support ventilator	Median AHI reduction from 37 to 10/h. Drawback of the set up : cannot be used to deliver BPAP because of insufficient trigger
Joseph et al. [11]	Israel	J Clin Sleep Med	Case series	5 children with OSA	2m-15 yrs	High Flow Nasal Cannula	One child used HFNC at home for 23 mo
Amaddeo et al. [12]	France	Sleep Med	Prospective study	8 CPAP non-compliant children, 6 Down syndrome, 1 Pierre Robin Sequence, 1 Pfeiffer	0.1 - 17.3 yrs	High Flow Nasal Cannula	Success in 5; mean compliance 7h10 ± 0.36 min/night Refusal (failure) in the 3 oldest patients with Down syndrome: 1 orthodontic treatment, 1 spontaneous improvement

				(one of the few studies with a clear definition of failed CPAP /non-adherence)			
Koncick et al. [13]	USA	Pediatric Pulmonology	Retrospective cohort on data from the Pediatric Health Information System (2007-2015)	3802 children with chronic respiratory failure (OSA excluded) discharged on NIV	< 21 yrs	Tracheotomy: 337 (8.9%) were transitioned to tracheotomy 58% had a neurologic disorders and 39% a NMD	Factors associated with a tracheotomy: younger age, anoxia/encephalopathy, quadriplegia
Diercks et al. [14]	USA	JAMA Otolaryngology Head Neck Surgery	Case series	6 Down syndrome adolescents not compliant to CPAP after upper airway surgery	12 - 18 yrs	Hypoglossal nerve stimulation	56% to 85% reduction in AHI and good use
Caloway et al. [15]	USA	Laryngoscope	Case series	20 Down syndrome non obese adolescents not compliant to CPAP after adenotonsillectomy	10 - 21 yrs	Hypoglossal nerve stimulation	Median reduction in AHI of 85%, good use

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Abbreviations: m: months, yrs: years, CPAP: continuous positive airway pressure, BPAP: bilevel positive airway pressure, NIV: noninvasive ventilation, NMD: neuromuscular disease, OSA: obstructive sleep apnea, MDO: mandibular distraction osteogenesis, NPT: nasopharyngeal tube, TLA: tongue-lip adhesion, PP: palatal plate, HFNC: high flow nasal cannula, AHI: apnea-hypopnea index, O2: oxygen, NICU: neonatal intensive care unit, BMI: body mass index.

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