Online Table S3.3: Initial and follow up settings for NIV

Author	Country	Journal	Type of study	Number of patients	Ages	NIV mode	NIV settings
Khan et al. [1]	UK	Arch Dis Child	Retrospective study	8 children with NMD: 4 congenital myopathy, 2 congenital muscular dystrophy, 2 rigid spine	6-13 yrs	All treated with NIV	IPAP 10-14 cmH_2O BUR 12-18/min
Nabatame et al. [2]	Japan	Brain Dev	Retrospective study	4 children juvenile Pompe disease	9-15 yrs	3 treated with NIV	IPAP 10-12 cmH_2O EPAP 3-4 cmH_2O
Mellies et al. [3]	Germany	Neuromuscul Disord	Prospective study	6 infants with SMA I and 1 with SMA II (+ 6 SMA controls without NIV)	6 – 11 yrs	7 treated with NIV	IPAP 10.5 cmH_2O EPAP 3.7 cmH_2O BUR 16/min
Fauroux et al. [4]	France	Crit Care Med	Prospective physiological study oesogastric pressure measures	8 children with CF	11-17 yrs	Comparison volume/targeted vs pressure-targeted mode	Similar efficacy of the 2 modes but greater decrease of the work of breathing when the patients adopted a controlled mode (+ greater subjective comfort by VAS)
Fauroux et al. [5]	France	Eur Respir J	Prospective physiological study: oesogastric pressure measures	10 children with CF	10-21 yrs	Pressure support	Better setting with oesogastric pressure measures: IPAP 12-20 cmH_2O (mean 16), high peak insp flow, sensitive inspiratory trigger, expiratory trigger 25-50%), less asynchrony

Abbreviations: yrs: years, NIV: noninvasive ventilation, ICU: intensive care unit, IPAP: inspiratory pressure, EPAP: expiratory pressure, BUR: back up rate, CF: cystic fibrosis, NMD: neuromuscular disease.

References

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4. Fauroux B, Pigeot J, Polkey MI, *et al.* In vivo physiologic comparison of two ventilators used for domiciliary ventilation in children with cystic fibrosis. *Crit Care Med* 2001; 29: 2097-2105.
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