Online Table S10: CPAP and NIV and quality of life for patients and families / caregivers

Author, year	Country	Journal	Type of study	Number of patients	Ages	QoL (family, child)
Nozoe et al. [1]	Brazil	Sleep & Breath	PSQI administered to mothers of Duchenne patients	32 caregiver mothers and 32 control mothers: 16 patients treated with NIV		Of the 16 NIV patients: 8 mothers had good sleep quality and 8 bad The longer the NIV use (months), the better mother sleep quality (=only sleep quality predictor)
Gonzalez et al. [2]	Spain	Eur J Pediatr	Cross-sectional study, interview (PedQoL)	41 patients (invasive and NIV) with 20 children treated with NIV	Median age of NIV patients: 10.1 yrs (7.1-13)	Lower QoL in patients and parents (compared to healthy children and their parents) Parents perceived lower QoL than the patients Importance of underlying disease (> 50% NMD) QoL scores were low in all age groups, mainly physical domain, scores are lower in children treated with NIV
Cadart et al. [3]	France	Acta Pediatr	Prospective	96 parents of 86 children with rare conditions and referred to a sleep lab for a P(S)G filled in questionnaires on anxiety, depression, family functioning, parents QoL, daytime sleepiness and sleep quality	0.3 - 18 yrs	27/86 children were treated with CPAP or NIV 19% of the parents of children on NIV/CPAP presented with moderate-to-severe anxiety; 8% presented with moderate-to-severe depression. The responses to all the questionnaires did not differ between parents of children treated or not treated with long term CPAP or NIV
Lynch et al. [4]	USA	Behavioral Sleep	OSA-specific QoL	42 youth-caregiver dyads: data from 25 youth-caregiver	8-16 yrs (exclusion 1. cognitive &	CPAP-adherent youth had decreased sleep disturbance + decreased caregiver concern

		Med	questionnaire given before and after 3 m of CPAP	dyads suitable for analysis	physical disabilities; 2. comorbid medical and neurological conditions; 3. on anti-psychotic drugs	No significantly different changes in overall QoL between CPAP adherent and non-adherent youths before CPAP and 3 m after initiation.
Redouane et al. [5]	Canada	Plos One	Single centre cross-sectional study on QoL of parents of children using G, IV or NIV, or both	39 using NIV, 8 using IV, 20 using G + NIV, and 8 G + IV	5-18 yrs	Parental perception of QoL was lower than those of parents with healthy children. QoL perception was lower in parents of children using G than IV/NIV; technologies do not have additive effect on QoL. No difference between children using IV or NIV: importance of underlying condition
Meltzer et al. [6]	USA	J Clin Sleep Med	Cross sectional prospective study comparing sleep patterns and Health releated QoL of parents of ventilated	42 families with ventilated children (30 IV and 12 NIV children) and 40 families of healthy children	4-17 yrs	Parents of children treated with IV or NIV showed significantly later bedtimes, shorter total sleep time, lower sleep efficiency. Many showed significant instability of their sleep, directly related to health related QoL.

			and healthy children			
Young et al. [7]	Australia	Neurology	Retrospective review of effect of NIV on clinical and QoL in children with severe NMD	14 children (SMA II, congenital myopathy, merosine deficient myopathy. myotonic dystrophy and Duchenne muscular dystrophy)	Median age 7.7 yrs (1.5-16 years)	Comparison before and after NIV initiation. Symptoms of daytime sleepiness (p=0.003) and headaches (p= 0.046) improved after initiation of NIV Sleep quality improved. QoL remained stable after NIV despite disease progression
Johannsen et al. [8]	Germany	Health and QoL outcomes	Questionnaire survey in 43 families of children with NMD	43 families returned questionnaire: 18 with ventilated children 25 non-ventilated children	Age of IV/NIV initiation 4.4 yrs ± 5.4	Compared to normative data, children with NMD and their families had a lower health-related-QoL and mental health. No additional negative influence on the overall health related QoL of ventilator use. IV/NIV per se is not responsible for the reduction of health related QoL and mental health
Baiardini I et al. [9]	Italy	J Child Neurology	Cross-sectional study using questionnaires (Child health questionnaire –parent form 30, Family	27 DMD children and their parents	Mean age of children 11.2 yrs Mean age of parents mean age 40 yrs	Out of 27 children, 4 were ventilator users. Children reported significantly lower scores than normative group in 10/15 children health questionnaire dimensions. Only the use of wheelchairs (p=0.02) and ventilators (p<0.001) was significantly associated to lower health related QoL. Family strain questionnaire scores were not influenced by children's characteristics.

			strain questionnaire			
Noyes J et al. [10]	UK	J Advanced Nursing	Cross sectional study using validated questionnaires	Comparison of ventilator dependent children reports of health related QoL with parents report and normative population 27 ventilated children (17 responded to questionnaires) 27 parents of ventilated children (15/27 on nocturnal NIV)	Age of children 4-18 yrs The underlying diagnosis were only classified into congenital/spinal injury/brain injury.	Ventilated patients report significantly lower overall health related QoL and significantly lower scores in all domains except about their friends, compared with healthy children and children with other chronic diseases. Parents reported significantly lower scores for their child's disease and relationships with friends. Positive correlation between children and parents in all areas apart from self-esteem and school. Parents are not necessarily accurate proxies for all aspects of their child's experience and perceptions.
Vuillerot C et al. [11]	France	J Adolescent Health	Cross sectional study using questionnaire to assess self perceived QoL correlating with impairment, disability	43 adolescents with NMD including DMD, SMA, congenital myopathy; congenital muscular dystrophy and fascioscapulo-humeral muscular dystrophy	mean age 13.8 ± 1.7 yrs	VSP-A (self-perceived health state in adolescents) assess perceived wellbeing in 10 domains. VSP-A scores in physically disabled adolescents were similar to those of non-disabled group with regard to vitality, body image, relationships with parents and friends, physical and psychological wellbeing. Adolescents with ventilatory support did not express lower scores than adolescents not requiring ventilatory support (p=0.39)

			and respiratory status			
Carneval e et al. [12]	Canada	Pediatr ics	A qualitative study using semistruct ured interview of 12 families with children on ventilatory support at home	12 families with children having: abnormal ventilatory control; NMD; spina bifida; craniofacial or airway abnormalities (4 tracheostomy invasive ventilation; 8 NIV) Fieldwork observations were carried out at home	Age not specified	6 principal themes were identified: 1. Confronting parental responsibilities – stressful for parents 2. Families seeking normality 3. Conflicting social values – parents feeling child's life devalued 4. Living in isolation 5. Voice of child often not heard 6. Questioning of moral order Despite difficulties described by families, they also reported deep enrichments and rewarding experiences that they could not imagine living without.

Abbreviations: m: months, yrs: years, QoL: quality of life, CPAP: continuous positive airway pressure, BPAP: bilevel positive airway pressure, NIV: noninvasive ventilation, IV: invasive ventilation, G: gastrostomy, PSQI: Pittsburgh Sleep Quality Index, PedQoL: Pediatric Quality of Life, NMD: neuromuscular disease, SMA: spinal muscular dystrophy, OSA: obstructive sleep apnea, P(S)G: poly(somno)graphy.

References

1. Nozoe KT, Polesel DN, Moreira GA, *et al.* Sleep quality of mother-caregivers of Duchenne muscular dystrophy patients. *Sleep Breath* 2016; 20: 129-134.
2. Gonzalez R, Bustinza A, Fernandez SN, *et al.* Quality of life in home-ventilated children and their families. *Eur J Pediatr* 2017; 176: 1307-1317.

3. Cadart M, De Sanctis L, Khirani S, *et al.* Parents of children referred to a sleep laboratory for disordered breathing reported anxiety, daytime sleepiness and poor sleep quality. *Acta Paediatrica* 2018; 107: 1253-1261.
4. Lynch MK, Elliott LC, Avis KT, *et al.* Quality of life in youth with obstructive sleep apnea syndrome (OSAS) treated with continuous positive airway pressure (CPAP) therapy. *Behav Sleep Med* 2019; 17: 238-245.
5. Redouane B, Cohen E, Stephens D, *et al.* Parental perceptions of quality of life in children on long-term ventilation at home as compared to enterostomy tubes. *PLoS One* 2016; 11: e0149999.
6. Meltzer LJ, Sanchez-Ortuno MJ, Edinger JD, *et al.* Sleep patterns, sleep instability, and health related quality of life in parents of ventilator-assisted children. *J Clin Sleep Med* 2015; 11: 251-258.
7. Young HK, Lowe A, Fitzgerald DA, *et al.* Outcome of noninvasive ventilation in children with neuromuscular disease. *Neurology* 2007; 68: 198-201.
8. Johannsen J, Fuhrmann L, Grolle B, *et al.* The impact of long-term ventilator-use on health-related quality of life and the mental health of children with neuromuscular diseases and their families: need for a revised perspective? *Health Qual Life Outcomes* 2020; 18: 219.
9. Baiardini I, Minetti C, Bonifacino S, *et al.* Quality of life in Duchenne muscular dystrophy: the subjective impact on children and parents. *J Child Neurol* 2011; 26: 707-713.
10. Noyes J. Comparison of ventilator-dependent child reports of health-related quality of life with parent reports and normative populations. *J Adv Nurs* 2007; 58: 1-10.
11. Vuillerot C, Hodgkinson I, Bissery A, *et al.* Self-perception of quality of life by adolescents with neuromuscular diseases. *J Adolesc Health* 2010; 46: 70-76.
12. Carnevale FA, Alexander E, Davis M, *et al.* Daily living with distress and enrichment: the moral experience of families with ventilator-assisted children at home. *Pediatrics International* 2006; 117: e48-e60.