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Parent carer quality of life and night-time attendance in non-ambulant youth with neuromuscular disorders

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TITLE: Parent carer quality of life and night-time attendance in non-ambulant youth with neuromuscular disorders.

ABSTRACT

Purpose: To describe and explore carer quality of life (QoL) and night-time attendance to their child in parents of non-ambulant youth with Neuromuscular Disorders.

Methods: A cross-sectional population-based, comprehensive survey including the Adult Carer QoL (AC-QoL) questionnaire, measures of social context and youths’ physical status. Associations between carer-QoL or frequency of parents’ night-time attendance with independent variables were explored using linear and logistic regression models, respectively.

Results: Parents’ perceived lower carer-QoL (mean 76.5/120, SD 18.5) when they attended to their child twice a night or more (n=17/35) and with shorter time since their child was prescribed non-invasive ventilation (NIV). Parental night-time attendance was not associated with youth’s actual use of NIV, but was more likely when youth required assistance to turn in bed, reported frequent sleep discomfort and had more severe joint contractures.

Conclusions: To optimise parent carer-QoL, interventions must address parents’ frequency of night-time attendance and youths’ sleep comfort.
Key Words:
Neuromuscular disorders; parent carer quality of life; youth; sleep; non-invasive ventilation

Abbreviations
AC-QoL - Adult Carer Quality of Life Questionnaire
NIV – Non-invasive ventilation
NMD – Neuromuscular Disorders
QoL - Quality of Life

Highlights:
In parents of non-ambulant youth with neuromuscular disorders:

- Lower carer quality of life was associated with more frequent night-time attendance.
- Almost half attend to their child twice a night or more.
- Nocturnal non-invasive ventilation use did not increase the likelihood of night-time attendance.
- Their child’s musculoskeletal discomfort was more likely to disrupt parents’ sleep.
1. **Introduction**

Youth with neuromuscular disorders (NMD) such as Duchenne muscular dystrophy (DMD), Spinal Muscular Atrophy and rarer dystrophies or myopathies live with global muscle weakness and progressive movement limitations. Youth who are unable to stand or walk are vulnerable to comorbidities in almost all body systems that limit survival. Best practice care can extend survival by decades, but is complex, involves multidisciplinary healthcare (including specialist doctors, nurses, rehabilitation therapists) and assistance from carers in almost all activities of daily living (e.g. transfers, toileting, dressing, eating). In particular, timely provision of non-invasive ventilation (NIV) has doubled survival, but technology dependence has added to the complexity of caregiving provided in the home. Though youth’s sleep may be improved with the support of NIV, the prospect of more complex care in the home may be daunting for both the parent and their child.

**Caregiver burden and quality of life**

Since children normally live with their parents, parents of youth growing up with disabilities by default become an invaluable resource as family caregivers, providing daily healthcare and practical support over and above usual parenting. A typical task of adolescence is to become physically independent in all daily activities with parents stepping back, but parents of non-ambulant youth with degenerative NMD’s are required to increasingly provide physical assistance as their child’s strength deteriorates. Caregiver burden in parents of children with NMD is linked with greater risk of depression, anxiety, reduced family function, elevated levels of stress, impact on work life and productivity and lower health-related quality of life (QoL). However, attempts to quantify caregiver burden reveal the subjective nature of this construct. Both negative and positive aspects of caregivers’ situations have been studied. Negative aspects and increased risk of caregiver burden are
linked to female carers, unemployed carers, a higher number of caregiving hours, greater financial distress, low educational attainment, less external agency care support, isolation and lack of choice in being a caregiver. Positive aspects are findings that include that caregiving is perceived as less burdensome when parents feel valued, adequately supported and experience reward and enrichment. Since the advent of technological support in the homes of youth with NMD, such as nocturnal NIV, the impact on parents’ night-time caregiving demands and carer QoL have received increasing attention.

Parents’ sleep quality

Parents who care for children dependent on technology report on average an hour less total sleep time, more disrupted sleep (a third more nocturnal awakenings measured by actigraphy) and poorer sleep quality (for example, longer time to fall asleep) than parents of typically developing children. Parents’ sleep problems negatively impact their physical health, increase symptoms of significant depression three-fold and impair executive functioning that can impact safety and ability to care for their family and disabled child during the day. In parents of young adults with DMD, poor sleep quality was linked with greater anxiety and poorer health related QoL. Yet their child’s use of NIV does not appear to be primary factor in parents’ poor sleep quality nor poorer health-related QoL. In parents of non-ambulant youth with NMD the frequency of night time attendance to their child and reasons for attendance are not well understood.

Youth factors

Beyond possibly attending to supportive technology such as NIV, parents of children with severe movement limitations endure sleep disruption to attend to their child’s multiple physical needs, such as to adjust body position or to manage concerns of breathing, pain and pressure care. In boys with DMD, movement limitation requiring assistance to turn was the
primary factor in parents’ reports of their son’s poor sleep quality\textsuperscript{16}. The same parents identified one in five boys with DMD as having a pathological total sleep score, on ratings of the Sleep Disturbance Scale for Children (SDSC), though no individual type of sleep disorder could be differentiated\textsuperscript{16}. Of particular interest in non-ambulant youth with NMD is an association of youth’s self-reported lower mental wellbeing with discomfort at night\textsuperscript{17}. A relationship of lower parent wellbeing with their youth’s lower wellbeing in typically developing youth is known\textsuperscript{18} but is to date underexplored in families with disability.

The current study

To enable continued deeper understanding of caregiver burden and carer QoL in parents of non-ambulant youth with NMD, there is a need to disentangle what goes on at night in families’ homes. The primary aim of this study was to describe carer QoL and parental attendance at night from the perspective of parents of non-ambulant youth with NMD. We expected to see a relationship where more frequent night-time attendance may be associated with lower parent carer QoL. A secondary aim was to explore the associations of parent carer QoL and parental attendance at night with social context and youth’s physical ability, respiratory and musculoskeletal status, and sleep comfort. Insights gained may identify priority areas on which to focus in order to support better outcomes in both parents and their youth.

2. Methods

2.1 Participants and recruitment

A cross sectional, population-based survey design study was conducted between September 2014 and July 2016 (the MyLifeMyVoice Teen NMD study; https://mylifemyvoice.org/). Eligible participants were non-ambulant youth with NMD aged 13 - 22 years and their parents living across Australia. Recruitment was by opt-in mailed invitations distributed by
third-parties that maintained confidentiality of their patient and member contact details. Nine patient organizations and five tertiary children’s hospital neuromuscular clinics sent invitations. Each site provided the first author (VT) with coded lists of individuals who were sent invitations, which were counted and cross-checked for duplication. This enabled an estimate of the number of eligible youth invited (Figure 1). Interested youth and their parents responded direct to the first author by e-mail, SMS, or by returning a stamped, addressed reply slip by mail. The first author telephoned respondents to screen for eligibility and sent the paper-based questionnaire via mail. Parents completed questionnaires independently and youth were offered physical assistance with completion if required. Parents and their youth provided written, informed consent returned together with the survey. Following return of the questionnaire, the first author conducted structured interviews with all families to minimise missing data and validate physical status data including assessment of physical ability, joint contracture and scoliosis. Interviews were conducted through home visits with families living in the author’s same state or via telephone or video call (Skype) with families living >600km from the author’s city. Ethics approval was provided by the human research ethics committees of all participating institutions.
2.2 Measures

**Parent Carer QoL**

Parent carer QoL was measured utilising the Adult Carer Quality of Life Questionnaire (AC-QoL)\(^9\). The scale comprises 40 items worded both positively and negatively, grouped into eight subscales: support for caring (items 1-5; example question: “I have all the practical support I need.”), caring choice (items 6-10; example question: “Caring stops me doing what I want to do.”), caring stress (items 11-15; example question: “I feel worn out as a result of caring.”), money matters (items 16-20; example question: “I feel satisfied with my financial situation.”), personal growth (items 21-25; example question: “I feel that I have become a better person by caring.”), sense of value (items 26-30; example question: “The person I look after makes me feel good about myself.”), ability to care (items 31-35; example question: “I can manage most situations with the person I care for.”) and carer satisfaction (items 36-40; example question: “I resent having to be a carer.”). Parents rated their experience “in the last
Scores are summed to a maximum of 120. According to the user manual, overall scores of 41 – 80/120 and subscale scores of 6 – 10/15 indicate mid-range reported QoL. Overall scores of 81+ /120 and subscale scores of 11+ / 15 indicate high reported QoL.

The AC-QoL additionally asks respondents to indicate the number of hours they spend caring in categories of 10 hours from “0 – 10 hours” to “> 71 hours” a week. Psychometric evaluation of the AC-QoL responses of parent carers of children with disabilities showed acceptable convergent and discriminant validity with measures of burden, resilience, satisfaction, physical and mental health. Preliminary evaluation of the AC-QoL provided evidence of acceptable internal consistency reliability and sensitivity to change.

**Social context**

Parents reported demographic information. Residence was reported by home location (state of residence, rural or metropolitan). Household composition was described by number of parents and children in the home. Parental employment status, country of birth and languages spoken other than English (to indicate family cultural background) were based on standardised questions published by the Australian Curriculum, Assessment and Reporting Authority. Frequency of external agency care assistance hours, including paid support from respite and personal carers, were recorded utilising the same question as a concurrent CARE-NMD study (personal communication, M Rodrigues, Muscular Dystrophy New Zealand, and Jan Kirschner, www.care-nmd.eu, Sept 2014). Responses were categorised as “daily”, “a few times a week” or “none”.

*two weeks* on 4-point Likert scale (“never = 0”, “some of the time = 1”, “a lot of the time = 2” and “always = 3”).
Youths’ physical status

Youths’ physical ability was measured utilising the Egen Klassification Scale Version 2 (EK2), the revised version following exploratory Rasch analysis\(^{23}\) of the original scale, which is a valid\(^ {24}\) and reliable\(^ {25}\) measurement of physical ability in non-ambulatory individuals with NMD. The scales’ 17 items describe physical ability in terms of ability to move the head, trunk and arms against gravity, stand, speak loudly and cough, chew and swallow, as well as assistance required with turning, transfer and meal intake. Each item is scored by choice of four or five discrete descriptors. The maximal score is 53. Higher scores indicate greater physical ability.

From responses to the EK2 scale Question 7 (related to ability to turn in bed with bedclothes), the need for assistance to turn in bed was dichotomised to assistance required or not.

Time since NIV prescription was calculated from the reported date of prescription to the date of questionnaire completion. Use of NIV was categorised into “not prescribed”, “trial or partial nocturnal” (trialled but not yet prescribed or used less than half the night or a few nights a week only) and “full nocturnal” (used every night for duration of sleep).

Scoliosis was described by the Cobb angle (degrees of spinal curvature deviating from the upright spine in the sagittal plane) reported in youth’s most recent Orthopaedic clinic review letter. Where families did not have a copy of the most recent review, the author followed up radiography results with their clinic with their consent. Scoliosis was categorised as “none”, “Cobb angle <40°” and “Cobb angle >40°” with or without spinal fusion.

In the absence of standardised descriptors of severity of joint contracture, novel descriptors relating to functional limitation were developed for this study. The number of severe joint contractures was totalled from responses to “very tight” descriptors of range of motion for
shoulder elevation, elbow flexion and extension, hip extension, knee flexion and extension,
and ankle dorsiflexion (for example, contracture at the right ankle joint was described as 0 =
“flexible in all directions”, 1 = “a bit tight: I can get normal shoes on and rest my right foot
on my wheelchair footplates”, 2 = “very tight: I can’t get normal shoes on and my right foot
hangs off my footplate/I can’t use footplates”).

Youth responded “Yes” or “No” to a question asking if they experienced pain at night.
Affirmative responses to experience of pain at night were utilised in analysis. Pain intensity
was measured utilising the Wong-Baker Faces Pain Rating Scale with ordinal response
options (0, 2, 4, 6, 8, 10) validated for use in children and shown to correlate well with the
continuous Visual Analog Scale (VAS).

Youths’ Sleep
A standardised measure of sleep quality valid in youth with physical disabilities could not be
sourced at the outset of this study. Sleep latency (time to fall asleep) and total sleep hours
were measured utilising the first two questions of the Sleep Disturbance Scale for Children
(SDSC). The complete scale was not utilised as items relying on the individual’s ability to
move (for example “The child has frequent twitching or jerking of legs while asleep or often
changes position during the night or kicks the covers off the bed”; “You have observed the
child sleepwalking.”) were not valid in a population of youth with NMD unable to move.

Novel questions were developed of parental attendance at night and youth sleep comfort with
Likert 5-point scale response options. A post-priori, pragmatic decision was made to
condense the number of categories to ensure sufficient cell size for meaningful analysis of
severe sleep issues as follows:

Responses to sleep latency were dichotomised to more or less than 60 minutes to fall asleep
and sleep duration to more or less than 8 hours’ total sleep.
Parents reported the number of times they attended to their child at night in categorical response options of “never”, “about once a week”, “about once every night”, “2-3 times a night” and “more than 3 times a night”, dichotomised for analysis into “once a night or less” and “twice a night or more”.

Youth self-reported their sleep comfort (“How often is comfort at night a problem?”) on a 5-point scale, reduced to three categories for analysis: “rarely” (including “never” and “almost never”), “sometimes” and “frequently” (including “often” and “almost always”).

Descriptors of positioning equipment used by youth at night in addition to their head pillow and blanket (for example, Symmetrisleep™ brackets, splints/braces and use of a specialist pressure mattress) were reduced to a binary variable describing sleep positioning equipment use or not.

### 2.3 Statistical analyses

Normally distributed data were expressed as means and standard deviations (SD), skewed data as medians and interquartile ranges (IQR), and categorical variables as frequencies and percentages.

The associations of parent AC-QoL and predictor variables were explored using linear regression models in two steps. Step 1 investigated associations between the independent variables and AC-QoL scores; the second step comprised a multivariable linear regression model including significant variables from those identified in Step 1.

The association between parental attendance at night (“once a night or less” versus “twice a night or more”) and predictor variables were analysed using binary logistic regression models, with model estimates and their 95% confidence intervals reported. A variable was considered significantly associated with the outcome if the Wald Chi-Square test of model
effects returned p < 0.05. All statistical analyses were calculated using SPSS® Statistics (Version 24, IBM Corp, New York: USA).

3. Results

Of the 289 families invited to participate, thirty-five parents (31 mothers, 4 fathers) and their child (median age 16.8 years, IQR 15.6 years – 18.5 years) returned completed questionnaires (Figure 1). Youth’s diagnoses included Duchenne muscular dystrophy (DMD) (n = 24, 69%, all male), Nemaline Rod Myopathy (n = 3, 9%, all female), Congenital Muscular Dystrophy (n = 6, 17%, 2 female) and Spinal Muscular Atrophy Type II (n = 2, 1 female).

Parent carer QoL

Parent’s ratings of carer QoL were normally distributed, with a mean AC-QoL score of 76.5/120 (SD 18.5, min 24 – max 114). One parent rated low (<40), 18 rated mid-range (41 – 80) and 16 rated high carer QoL (>81).

Parent carer QoL described by AC-QoL subscales is presented in Table 1. Parents rated perceived carer QoL lowest in the subdomain ‘support for caring’, though the mean fell just within the mid-range (6 – 10) of this scale (mean 7.3/15 SD 3.4). Parents rated high perceived QoL (11+) in the subdomains ‘sense of value’ (11.9/15 SD 3.7), ‘ability to care’ (11.1/15 SD 2.8) and ‘carer satisfaction’ (11.1/15 SD 2.6).
Table 1 Univariable effects of parental attendance at night and parent carer QoL (AC-QoL) subscales (n = 35)

<table>
<thead>
<tr>
<th>Parent Carer AC-QoL Mean (SD)</th>
<th>Parent attendance at night</th>
<th>Time since NIV prescription</th>
<th>β coefficients (95% CI)</th>
<th>β coefficients (95% CI)</th>
<th>p³</th>
<th>p³</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>≤ once a night n = 18</td>
<td>≥ twice a night n = 17</td>
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</tr>
<tr>
<td><strong>1 Support for caring</strong></td>
<td>7.3 (3.4)</td>
<td>0.0 (reference)</td>
<td>-2.10</td>
<td>(-4.20 to -0.05)</td>
<td>0.049</td>
<td>0.09</td>
</tr>
<tr>
<td></td>
<td>(95% CI)</td>
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<tr>
<td><strong>2 Caring choice</strong></td>
<td>8.6 (3.6)</td>
<td>0.0 (reference)</td>
<td>-2.37</td>
<td>(-4.56 to -0.18)</td>
<td>0.034</td>
<td>0.04</td>
</tr>
<tr>
<td></td>
<td>(95% CI)</td>
<td></td>
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<tr>
<td><strong>3 Caring stress</strong></td>
<td>9.4 (3.1)</td>
<td>0.0 (reference)</td>
<td>-3.24</td>
<td>(-4.95 to -1.53)</td>
<td>&lt;0.001</td>
<td>0.13</td>
</tr>
<tr>
<td></td>
<td>(95% CI)</td>
<td></td>
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<tr>
<td><strong>4 Money Matters</strong></td>
<td>8.4 (4.4)</td>
<td>0.0 (reference)</td>
<td>-4.38</td>
<td>(-6.84 to -1.92)</td>
<td>&lt;0.001</td>
<td>0.10</td>
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<tr>
<td></td>
<td>(95% CI)</td>
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<tr>
<td><strong>5 Personal Growth</strong></td>
<td>8.7 (3.7)</td>
<td>0.0 (reference)</td>
<td>-1.04</td>
<td>(-3.46 to 1.37)</td>
<td>0.398</td>
<td>0.46</td>
</tr>
<tr>
<td></td>
<td>(95% CI)</td>
<td></td>
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<td></td>
<td></td>
<td>(0.21 to 0.71)</td>
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<tr>
<td><strong>6 Sense of Value</strong></td>
<td>11.9 (3.7)</td>
<td>0.0 (reference)</td>
<td>-2.70</td>
<td>(-4.91 to -0.48)</td>
<td>0.017</td>
<td>0.27</td>
</tr>
<tr>
<td></td>
<td>(95% CI)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>(-0.06 to 0.60)</td>
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<tr>
<td><strong>7 Ability to care</strong></td>
<td>11.1 (2.8)</td>
<td>0.0 (reference)</td>
<td>-0.89</td>
<td>(-2.61 to 1.01)</td>
<td>0.387</td>
<td>0.18</td>
</tr>
<tr>
<td></td>
<td>(95% CI)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>(-0.07 to 0.43)</td>
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<td></td>
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</tr>
<tr>
<td><strong>8 Carer satisfaction</strong></td>
<td>11.1 (2.6)</td>
<td>0.0 (reference)</td>
<td>-1.65</td>
<td>(-3.25 to -0.06)</td>
<td>0.042</td>
<td>0.17</td>
</tr>
<tr>
<td></td>
<td>(95% CI)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>(-0.02 to 0.35)</td>
</tr>
</tbody>
</table>

Bolded numbers indicate significant association (95% CI excluding zero); ¹mean score out of total 15 for subscale; ²Wald-Chi Square Test of Model Effects value

Compared with a large group of Italian parents of slightly younger children with various physical disabilities {Negri, 2019 #5110, global ratings of carer QoL were lower in this Australian sample (AC-QoL mean 76.5/120, SD 18.5 compared with mean 85.9/120, SD 15.3). Greatest mean differences between the current sample and the Italian sample were in domains of caring choice (Australian parents’ mean 2.7 points lower) and caring stress (Australian parents’ mean 2.2 lower) (see APPENDIX).
Social context

Diverse social contexts were represented (Table 2), though a greater number of families were two parent households (89%) with the responding parent employed (69%) and university educated (51%). Nearly half of responding parents (n=16, 46%) reported spending more than 71 hours per week caring for their child. Just under a third of families (n=11, 31%) reported receiving daily external agency assistance with their child’s care. Youth’s physical ability as per EK2 scale scores was normally distributed (mean 33.5/53, SD 8.4, range 15 – 47). All required assistance to stand and none could take steps. Twenty-three youth (66%) required assistance to turn in bed with bedclothes. Twenty-three youth (66%) had been prescribed NIV, with time since prescription ranging from 1 month to 14.6 years.
### Table 2: Uni- and multivariable associations of parent carer QoL (AC-QoL) with social context, youths’ physical status and parental attendance at night

<table>
<thead>
<tr>
<th>Predictor Variable</th>
<th>N = 36</th>
<th>Parent Carer AC-QoL (mean: 76.5/120 SD: 18.5)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td><strong>UNIVARIABLE ASSOCIATION</strong></td>
</tr>
<tr>
<td></td>
<td>n (%) / Mean (SD)</td>
<td>β coefficients (95% CI)</td>
</tr>
<tr>
<td><strong>Household composition</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Two parents</td>
<td>31 (85%)</td>
<td>0.0 (reference)</td>
</tr>
<tr>
<td>Single parent</td>
<td>4 (11%)</td>
<td>6.48 (-12.44 to 25.39)</td>
</tr>
<tr>
<td><strong>Number of siblings</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2 (SD 1) (range 0–4)</td>
<td>2.40 (-9.26 to 1.26)</td>
<td>0.136</td>
</tr>
<tr>
<td><strong>Responding parent employed</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>24 (65%)</td>
<td>0.0 (reference)</td>
</tr>
<tr>
<td>No</td>
<td>11 (31%)</td>
<td>-3.27 (-16.27 to 9.73)</td>
</tr>
<tr>
<td><strong>Responding parent’s education</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>University trained</td>
<td>18 (51%)</td>
<td>0.0 (reference)</td>
</tr>
<tr>
<td>Vocation trained</td>
<td>9 (26%)</td>
<td>-15.720 (-29.39 to -2.05)</td>
</tr>
<tr>
<td>School only</td>
<td>8 (23%)</td>
<td>-5.58 (-19.81 to 8.54)</td>
</tr>
<tr>
<td><strong>Responding parents’ care hours</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>&gt; 71 hrs/week</td>
<td>16 (46%)</td>
<td>0.0 (reference)</td>
</tr>
<tr>
<td>31–70 hrs/week</td>
<td>10 (29%)</td>
<td>-0.73 (-14.53 to 13.08)</td>
</tr>
<tr>
<td>0 – 30 hrs/week</td>
<td>9 (26%)</td>
<td>12.04 (-22.22 to 26.31)</td>
</tr>
<tr>
<td><strong>External agency care assistance hours</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Daily</td>
<td>11 (31%)</td>
<td>0.0 (reference)</td>
</tr>
<tr>
<td>A few times a week</td>
<td>15 (43%)</td>
<td>-11.52 (-23.73 to 0.69)</td>
</tr>
<tr>
<td>None</td>
<td>9 (26%)</td>
<td>11.55 (-21.71 to 25.48)</td>
</tr>
<tr>
<td><strong>Even Classification 2 score of youth’s physical ability</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>33.3 (SD 8.6)</td>
<td>0.71 (0.03 - 1.38)</td>
<td>0.040</td>
</tr>
<tr>
<td>Time since NIV prescription (n=23)</td>
<td>4.1 years (SD 4.4 years)</td>
<td>1.42 (0.12 to 2.72)</td>
</tr>
<tr>
<td>Parental attendance at night</td>
<td></td>
<td></td>
</tr>
<tr>
<td>≤ once a night</td>
<td>18 (51%)</td>
<td>0.0 (reference)</td>
</tr>
<tr>
<td>≥ twice a night</td>
<td>17 (49%)</td>
<td>-18.27 (-28.77 to -7.77)</td>
</tr>
</tbody>
</table>

Bolded numbers indicate significant association (95% CI excluding zero); \(^1\)Wald-Chi Square Test p value outcome for the hypothesis \(\beta = 0\); \(^2\)Wald-Chi Square Test for Model Effects; \(^3\)Higher scores indicate greater physical ability. Maximum score is 53.
Univariable effects on parent carer QoL are shown in Table 2. Parents with university education rated their AC-QoL scores on average 16 points higher than parents with vocational training (coefficient 15.72; 95% CI 2.05 to 29.39, p = 0.024). Though parents who received daily external agency care assistance rated higher carer QoL than parents who received care a few times a week (coefficient -11.52; 95% CI -23.73 to 0.69, p = 0.064), the former parents’ ratings were lower than those who reported no agency care (coefficient 11.66; 95% CI -2.71 to 25.48, p = 0.098). Youth’s greater physical ability was associated with a less than one point higher parent AC-QoL score (coefficient 0.71; 95% CI 0.03 to 1.38, p = 0.040). A longer time since NIV prescription was associated with greater parental AC-QoL (coefficient 1.42; 95% CI 0.12 to 2.72, p = 0.033). Parents who attended their child once a night or less rated their AC-QoL scores on average 18 points higher compared to parents who attended their child twice a night or more (coefficient 18.27; 95% CI 7.77 to 28.77; p = 0.001).

Combining all significant variables identified in the univariable models, greater parent carer QoL remained independently associated with a longer time since NIV prescription and with parental attendance once a night or less (Table 2). The latter two variables were thus examined in more detail with AC-QoL subdomains (Table 1). A longer time since NIV prescription was associated with greater AC-QoL subdomain ‘personal growth’ (coefficient 0.46; 95% CI 0.21 to 0.71). Attending to their child twice a night or more was associated with lower carer QoL in six of the eight subdomains, with the greatest negative deviation in subdomains ‘money matters’ (coefficient -4.38; 95% CI -6.84 to -1.92; p < 0.001) and ‘perceived caring stress’ (coefficient -3.24; 95% CI -4.95 to -1.53; p < 0.001).
Youths’ physical status

Youths’ assistance required to turn in bed, respiratory and musculoskeletal status, and sleep comfort are shown in Table 3. Sixteen of the 23 youth prescribed NIV consistently used their device every night and seven reported trial or partial use. Parents of four youth consistently using NIV reported never attending to their child at night.

All youth reported joint contractures, with 27 (77%) reporting severe contractures in one joint (n = 10), two joints (n = 9) and three or more joints (n = 8). More than half of youth reported experiencing pain at night (n = 19, 54%), with intensity ranging from 2 to 8 out of 10.

Almost three quarters of youth (n = 26, 74%) used equipment in bed to position their body during sleep, including 9 (23%) who used lower limb splints (three at the knees and six at the ankles) and 11 (28%) who used additional pillows at the knees. Nineteen (49%) used an air alternating pressure care mattress.

One third of youth (n = 13, 37%) reported experiencing sleep discomfort “often” or “almost always”, including 12 youth requiring assistance to turn, eight of the 19 youth with specialist pressure mattresses, eight of the 16 full-nocturnal NIV users, seven youth who experienced pain every night, and four of the six youth who also had electric turning beds.
Table 3 Youths’ physical status and odds ratios (OR) of parental attendance “twice a night or more” compared with “once a night or less”

<table>
<thead>
<tr>
<th>Youth variable</th>
<th>Group</th>
<th>n (%)</th>
<th>count</th>
<th>count</th>
<th>OR</th>
<th>95% CI</th>
<th>p^1</th>
</tr>
</thead>
<tbody>
<tr>
<td>Assistance required to turn</td>
<td>No</td>
<td>12 (34%)</td>
<td>11</td>
<td>1</td>
<td>1.0</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Time since NIV prescription (n=23)</td>
<td>Yes</td>
<td>23 (66%)</td>
<td>7</td>
<td>16</td>
<td>25.1</td>
<td>2.7 – 234.2</td>
<td>0.005</td>
</tr>
<tr>
<td>NIV use</td>
<td>Not prescribed</td>
<td>12 (34%)</td>
<td>8</td>
<td>4</td>
<td>1.0</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Trial / partial use</td>
<td>7 (20%)</td>
<td>3</td>
<td>4</td>
<td>1.0</td>
<td>0.1 – 2.6</td>
<td>0.375</td>
</tr>
<tr>
<td></td>
<td>Full nocturnal use</td>
<td>16 (48%)</td>
<td>7</td>
<td>9</td>
<td>2.9</td>
<td>0.1 – 1.8</td>
<td>0.389</td>
</tr>
<tr>
<td>Sleep comfort problems</td>
<td>Never/almost never</td>
<td>14 (40%)</td>
<td>12</td>
<td>2</td>
<td>1.0</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Sometimes</td>
<td>8 (23%)</td>
<td>4</td>
<td>4</td>
<td>5.5</td>
<td>0.7 – 42.6</td>
<td>0.103</td>
</tr>
<tr>
<td></td>
<td>Often or almost always</td>
<td>13 (37%)</td>
<td>2</td>
<td>11</td>
<td>33.0</td>
<td>3.9 – 276.0</td>
<td>0.001</td>
</tr>
<tr>
<td>Deformity – Spinal scoliosis</td>
<td>No scoliosis</td>
<td>10 (29%)</td>
<td>7</td>
<td>3</td>
<td>1.0</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>&lt;40° Cobb angle</td>
<td>19 (54%)</td>
<td>8</td>
<td>11</td>
<td>0.7</td>
<td>0.1 – 4.6</td>
<td>0.735</td>
</tr>
<tr>
<td></td>
<td>&gt;40° Cobb angle</td>
<td>5 (17%)</td>
<td>3</td>
<td>3</td>
<td>2.3</td>
<td>0.3 – 19.0</td>
<td>0.428</td>
</tr>
<tr>
<td>Joint contractures</td>
<td>Median 1 [IQR 0 - 2]; min 0, max 9</td>
<td>1.2</td>
<td>1.1 – 1.2</td>
<td>&lt;0.001</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pain at night</td>
<td>No</td>
<td>16 (46%)</td>
<td>10</td>
<td>6</td>
<td>1.0</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Yes</td>
<td>19 (54%)</td>
<td>8</td>
<td>11</td>
<td>2.3</td>
<td>0.6 – 8.9</td>
<td>0.233</td>
</tr>
<tr>
<td>Sleep positioning equipment</td>
<td>No</td>
<td>9 (26%)</td>
<td>6</td>
<td>3</td>
<td>1.0</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Yes</td>
<td>26 (74%)</td>
<td>12</td>
<td>14</td>
<td>2.3</td>
<td>0.5 – 11.4</td>
<td>0.295</td>
</tr>
<tr>
<td>Sleep latency</td>
<td>&lt;60 min</td>
<td>31 (89%)</td>
<td>17</td>
<td>14</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>&gt;60 min</td>
<td>4 (11%)</td>
<td>1</td>
<td>3</td>
<td>5.5</td>
<td>0.3 – 39.0</td>
<td>0.285</td>
</tr>
<tr>
<td>Sleep duration</td>
<td>≥ 8 hours per night</td>
<td>24 (69%)</td>
<td>14</td>
<td>10</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>&lt; 8 hours per night</td>
<td>11 (31%)</td>
<td>4</td>
<td>7</td>
<td>2.5</td>
<td>0.6 – 10.7</td>
<td>0.233</td>
</tr>
</tbody>
</table>

Bolded numbers indicate significant association (p < 0.05). P value of Wald-Chi Square hypothesis test; NIV – Non Invasive Ventilation.
Table 3 shows the frequency and odds ratios for parents attending to their child twice a night or more compared with parents attending once a night or less. Compared with youth who did not require assistance to turn in bed, the odds of a parent attending twice a night or more were 25 times higher when their child required assistance to turn in bed (95% CI 2.7 to 234.2). Compared with youth who reported no sleep discomfort, the odds of a parent attending twice a night or more were 33 times higher when their child reported sleep discomfort “often or almost always” (95% CI 3.9 to 276.0). The odds of parents attending to their child twice a night or more were significantly higher when their child had more joint contractures (OR 1.2, 95% CI 1.1 to 1.2). No association was detected between parental attendance at night and NIV use, time since NIV prescription, scoliosis, pain at night, sleep positioning equipment, sleep latency nor sleep duration.

4. Discussion

This study aimed to describe and explore associations of carer QoL and attendance to their child at night in parents of non-ambulant youth with NMD. Insights gained provide some direction to find ways to better support both parents’ and their youth’s outcomes. Parents reported on average mid-range carer QoL measured using the AC-QoL tool. Social contexts, specifically lower parental education, caregiving more than 71 hours per week and inconsistent external agency care were linked with lower carer QoL. Of particular concern was the finding that half of youths’ parents attended to their child twice a night or more and that this factor remained independently associated with lower parent carer QoL. Our findings are in keeping with previous studies identifying caregiver burden linked to poor sleep in parents of children with complex disability. Parental attendance at night appeared unrelated to their child’s use of NIV. The higher likelihood of parental attendance when their child reported frequent sleep comfort problems and required assistance to turn in bed
highlights how youths’ movement limitations other than the need for respiratory support are
factors impacting parents’ night-time caregiver burden.

Parent Carer QoL

Parents rated global carer QoL in the mid-range. Two similar studies, one of 10 parent
caregivers of non-ambulant young adults with NMD\textsuperscript{13} and one of 42 parents of children
dependent on medical technology\textsuperscript{10} also reported overall health related QoL comparable to
the average population (on the SF-36 and SF-12 respectively). In both the latter populations,
subscale analysis identified lower than average mental health. A similar pattern was found in
our study; recent psychometric evaluation of the AC QoL\textsuperscript{20} found correlation of SF-36
mental health subscales with AC QoL subscales caring stress and caring choice. Lower
ratings in caring choice, caring stress and especially support for caring in the current sample
compared with Italian parents of children with disability\textsuperscript{20} indicate that our sample
experienced compromised wellbeing and caregiver burden\textsuperscript{7}. There may be differences in
social context, cultural expectations of caregiving and service provision support in the
Australian compared with the European context.

Social context

Good social support, higher education and higher income enable less perceived caregiver
burden and greater QoL in parents of children with NMD\textsuperscript{6} and parents of children on
nocturnal NIV\textsuperscript{10}. Our study confirms the opposite alongside previously reported studies, that
is, lower parental education\textsuperscript{10}, a greater number of parental hours spent caring per week\textsuperscript{7} and
less external agency care assistance\textsuperscript{20} also negatively impacted carer QoL in our sample.

Parents who have inconsistent care may have more difficulty juggling the demands of home
and employment. Parents with daily or no care may have a more predictable routine, or social
or personal resources enabling family functioning\textsuperscript{30} and resilience not captured in this study.
Notwithstanding, the markedly lower rating by our parents in the domain support for caring in comparison to Italian parents may be a reflection of different service provision. The Italian families were serviced by the same organisation\textsuperscript{20}, whereas Australian families are currently navigating a changing landscape of in-home support though a shift to the National Disability Insurance Scheme (NDIS), a national scheme purchasing local services based on individual service plans. The vast geographical dispersion and inconsistent funding to individual families are a challenge to equitable service provision\textsuperscript{31} and advocacy for greater funding and transparent planning processes is ongoing\textsuperscript{32}.

Less leisure time and less money increases carer stress\textsuperscript{7}, which is also associated with parents’ greater anxiety, depression\textsuperscript{14} and poorer sleep\textsuperscript{10, 14}. Greatest negative deviation in AC-QoL subdomains money matters and carer stress associated with frequent night-time assistance reinforces the circular association of parent’s poor sleep quality and compromised mental health\textsuperscript{33}. In combination with the social contextual factors affirmed above, the finding that more frequent attendance at night was negatively associated with almost all subscale measures of carer QoL affirms the need for consistent carer support widely called for \textsuperscript{10, 11, 29, 34}. Resources must be directed to alleviating the burden of night-time caregiving.

**Reasons for attending at night**

Our study affirms that the addition of medical technology such as NIV to home care does not necessarily require more night-time care in the long term, but does require a period of adjustment during which families need more support. Use of NIV can be hampered by youth’s fears of technology dependence\textsuperscript{35} and families’ difficulties managing the device at home\textsuperscript{4}. Initial impact on parental QoL has previously found parental waking due to anxiety even when their young person’s sleep is not disrupted\textsuperscript{11}. Interruption to parents’ deep restorative sleep cycle will have greatest impact on parent’s sleep quality and wellbeing\textsuperscript{10}. 
Initial impact on parental waking may include attending to device alarming due to set up difficulties such as poor mask or other interface fit, or device asynchrony to the young person’s breathing pattern. Adequate nocturnal NIV set up has been shown to optimise youth’s sleep and families adapt to NIV over time, also predicted by family functioning at the outset. Longitudinal studies are needed to explore cost-benefit implications of initiating NIV through specialist services and providing overnight, in-home care at least in the initial period to optimise device set up and parents’ wellbeing.

Youth’s physical status

Findings of our study add to understanding of why parents attend to their child at night for reasons other than respiratory support, directing attention to musculoskeletal comfort. In keeping with previous studies, most of the youth whose parent attended to them frequently required assistance to turn in bed. We found an association of parental attendance at night with youth’s self-reported sleep discomfort and more severe joint contracture, but not with our crude measure of pain. Previous studies have linked parental night-time attendance to their child’s pain, reported by the parent. Our measure may not have been sensitive enough. Alternatively, youth may not necessarily equate discomfort with pain, but may experience anxiety with discomfort and the inability to move, impacting sleep and calls for parental assistance. Mindful that typically developing individuals move 20–40 times or more a night, mostly without waking, there is no doubt that sleep comfort needs addressing. Joint contractures may cause discomfort, yet whether supine sleep positioning equipment and splints that aim to manage deformity relieve or add to sleep discomfort is underexplored. For individuals with some movement, sleep positioning equipment may limit independent ability for repositioning. The high proportion of frequent sleep discomfort and parental
attendance reported in youth who also used pressure mattresses and turning beds suggests that either that these assistive technologies require further development, or parents need physical help overnight.

Although new drug developments increase hope for some maintenance of movement ability\textsuperscript{[41]}, rehabilitation care still needs to find ways to optimise movement over 24 hours a day in individuals with NMD with no movement ability. Further investigation is warranted of youths’ musculoskeletal management overnight (for example how sleep positioning aides are used) as well as physical activity during the day (for example electric wheelchair sports, swimming or sit-stand wheelchairs) that may improve flexibility and mental wellbeing.

4.1 Strengths and limitations

The strengths of this study are the examination of parent as well as youths’ self-reported experiences of night-time care in a sample homogenous by age and functional ability - all were non-ambulant wheelchair users. The sample was diverse by service provision, since clinic funding, organisation and disability care provision varies in centres across Australia\textsuperscript{[31, 32]}. The findings of this study may apply to populations of non-ambulant youth with other physical disabilities and severe movement limitations cared for in different health service contexts. Whilst the sample included youth from different social contexts and family structures, the majority represent two-parent households with tertiary educated parents who are employed. Thus, generalisability may be limited to those within similar social contexts.

The current study’s sample size was small and further investigation will be helpful to affirm or refute whether the physical and possible psychological factors identified in this study impact night-time care in similar populations of non-ambulant youth\textsuperscript{[42]}.

This study has limitations. (1) Cross sectional data cannot infer causative relationships, only associations. Risk of type two error is increased with small sample size and other associations
may be found or negated in larger samples. (2) Opt-in recruitment required by the human research ethics committees for this study may have favoured participation of families who had capacity to respond to a research invitation and/or were highly motivated to participate. That is, either families coping comparatively well with their burden of care or families who felt their QoL was more impacted may be over-represented. Future study should consider opt-out recruitment strategies to give more youth and their families opportunity to consider study participation. (3) Relationships between constructs may be inflated or distorted by the effect of common method variance due to utilisation of a single data sourced by a questionnaire survey. Additionally, questionnaire measures of sleep quality such as the SDSC was limited by such tool’s questions related to independent movement and lack of validation in youth with physical disabilities. Future study will benefit from use of a greater variety of data collection tools and standardised assessment of sleep quality relevant to and validated in carers of youth with complex disability, including actigraphy and the recently developed HOST questionnaire. (4) The single descriptor of frequency of parental attendance at night did not capture the full possible extent of interruption to parents’ sleep quality or sleep cycle. Future studies should also comprehensively capture reasons parents attend to their child, the time of night and length of the attendance, and measures of the parent’s deep restorative sleep cycle pattern.

5. Conclusions

Measurement of parent carer QoL using the AC-QoL tool with subscale analysis provided evidence of caregiver burden in parents of youth with NMD complementary to previous studies. Findings of association of lower parent carer QoL with more frequent night-time attendance are not new. Our findings reinforce previous studies’ calls for monitoring of
parents’ stress, depression, anxiety\textsuperscript{10} and sleep quality\textsuperscript{11} and to provide adequate economic and social support resources to optimise parent carer QoL\textsuperscript{48}.

Importantly, this study adds detail of frequency of parental attendance at night in combination with youths’ self-reported possible reasons for disrupted nights in the parent-child dyad. This study reassures that the longer the young person uses nocturnal NIV, respiratory support is not associated with their parents’ night-time attendance. The findings indicate that more attention needs to be paid to the impact of youth’s movement limitations over the 24-hour cycle. Frequency of and reasons for nightly attendance to their child will be useful measures to add to future study investigating caregiver burden in parents of children with complex disability. To optimise parent carer QoL, assessment of youths’ sleep comfort and adequate interventions and support to minimise parents’ frequency of night-time attendance should be an integral part of care provided to families of non-ambulant youth with NMD.
Acknowledgements

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Disclosure of interests: The authors report no conflict of interest.
References


647
648
649
650
651
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653
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655
656
657
658
**APPENDIX**

**Table**  Comparison of carer QoL (AC-QoL) in current sample with Italian parents\(^1\)  
(higher scores indicate greater carer QoL)

<table>
<thead>
<tr>
<th>Child’s mean age (SD)</th>
<th>Current Australian sample (n = 35)</th>
<th>Italian sample (n = 591)</th>
</tr>
</thead>
<tbody>
<tr>
<td>17.5 years old (2.4)</td>
<td>11.1 years old (3.31)</td>
<td></td>
</tr>
</tbody>
</table>

**Child’s functional ability**  
- Non-ambulant, physical disability
- Range of disease severity,  
  “High” severity in n=259 (44%).  
  (mental & behavioural disorders in 60.7%;  
  nervous system diseases in 24.4%)  
- No correlation found of parent AC-QoL score and child disease severity.

<table>
<thead>
<tr>
<th>Total AC-QoL score Mean / 120 (SD)</th>
<th>76.5 (18.5)</th>
<th>85.9 (15.3)</th>
</tr>
</thead>
</table>

<table>
<thead>
<tr>
<th>Subscale scores</th>
<th>Mean / 15 (SD)</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>1 Support for caring</td>
<td>7.3 (3.4)</td>
<td>9.3 (3.2)</td>
</tr>
<tr>
<td>2 Caring choice</td>
<td>8.6 (3.6)</td>
<td>11.3 (3.3)</td>
</tr>
<tr>
<td>3 Caring stress</td>
<td>9.4 (3.1)</td>
<td>11.6 (2.9)</td>
</tr>
<tr>
<td>4 Money Matters</td>
<td>8.4 (4.4)</td>
<td>8.3 (3.3)</td>
</tr>
<tr>
<td>5 Personal Growth</td>
<td>8.7 (3.7)</td>
<td>10.3 (3.2)</td>
</tr>
<tr>
<td>6 Sense of Value</td>
<td>11.9 (3.7)</td>
<td>12.7 (2.8)</td>
</tr>
<tr>
<td>7 Ability to care</td>
<td>11.1 (2.8)</td>
<td>11.3 (2.6)</td>
</tr>
<tr>
<td>8 Carer satisfaction</td>
<td>11.1 (2.6)</td>
<td>11.2 (2.9)</td>
</tr>
</tbody>
</table>