Neuromuscular Disorders 20 (2010) 458–463

Contents lists available at ScienceDirect

Neuromuscular Disorders

journal homepage: www.elsevier.com/locate/nmd

Sleep and well-being in young men with neuromuscular disorders receiving non-invasive ventilation and their carers

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ARTICLE INFO

Article history:
Received 16 December 2009
Received in revised form 10 May 2010
Accepted 24 May 2010

Keywords:
Neuromuscular disorder
Ventilation
Well-being

ABSTRACT

Nocturnal ventilation has improved the physical status and life span of childhood neuromuscular disorders: the purpose of this study was to assess the implications for sleep and well-being in patients and carers. Ten young men (age range 12–25 years) with neuromuscular disorders on assisted ventilation and/or their main carers completed questionnaires on sleep quality, physical and psychological well-being, family burden and function. Both patients and parents expressed satisfaction with ventilation treatment. Compared to standardised values patients reported reduced sleep quality, but their mental health was not substantially affected. Poor sleep quality in carers – but not in patients – was significantly associated with risk for emotional (anxiety and depressive) disorders, reduced physical/emotional health, family burden and difficulty. We conclude that patients were generally well adapted psychologically, but sleep quality was poor and in carers this was linked to reduced well-being and family burden.

1. Introduction

Respiratory impairment is a major threat to life for young people with neuromuscular disorders: most deaths in Duchenne Muscular Dystrophy (DMD) are due to pulmonary infections and respiratory insufficiency [1]. Advances in treatment include the use of non-invasive nocturnal ventilation. This aims to decrease hypercapnia, and improve vital lung capacity as well as daytime sleepiness, fatigue and alertness [2]. Evidence suggests that it leads to positive changes in gas exchange, maintained for over 5 years without decline, and that patients can derive palliation of symptoms and improved quality of life as well as prolongation of life [3,4]. Ventilation has revolutionised care and prolonged life in boys with Duchenne Muscular Dystrophy; the mean age of death was 14.4 years in the 1960's but for those ventilated since 1990 it rose to 25.3 years [5]. Current survival figures from our own cohort of ventilated DMD individuals suggest that the mean age of death is between 29 and 30 years (Simonds, personal observation).

Non-invasive ventilation can be provided at home overnight; this is more convenient than invasive ventilation and has fewer adverse effects on speech, appearance and comfort. Most patients prefer it because they can remain at home longer and require less professional intervention and fewer hospital admissions [6,7]. While physical function remains compromised health perceptions in patients with neuromuscular disorders on ventilation is generally positive and has been found to be superior to that of older patients with chronic obstructive pulmonary disease [8–11].

Nevertheless, nasal ventilation takes place predominantly overnight and has the potential to affect sleep and, consequently, well-being in patients and carers. It requires high levels of care and attention from parents and every aspect of family life can be highly complicated [12]. It may stretch family coping [4], especially if marred by poorly directed social and medical support [13]. However, little attention has been directed at documenting sleep quality and coping capacity in patients and families.

In this study we aimed to examine sleep and well-being in young men with neuromuscular conditions on assisted ventilation and their main carers. We hypothesized that ventilation would be associated with sleep problems and that the latter would be linked to patient and carer well-being.

2. Patients and methods

2.1. Participants

The sample consisted of carers and patients with neuromuscular disorder aged 12 to 25 years receiving nocturnal ventilation at

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doi:10.1016/j.nmd.2010.05.011

the Brompton Hospital, a specialist respiratory centre covering a wide geographical area in south west England, stratified by length of time on nocturnal ventilation (under one year, 1–5 years and over 5 years). Ethical approval for the study was given by Brompton, Harefield & NHLI Ethics Committee.

Main carers and patients were requested to complete and return postal questionnaires and were also invited to take part in a semi-structured interview to provide their views on the treatment and general coping.

2.2. Measures

A questionnaire constructed for the study obtained information from patients and main carers on family socioeconomic status, family composition, education, the health status of the patient and main carer, their views about ventilation treatment and other family supports available.

Sleep was assessed by patients and carers completing the *Pittsburgh Sleep Quality Index* [14]. This self-report 19-item tool measures sleep quality and disturbance retrospectively over a 1-month period and generates seven component scores, related to sleep duration and latency, frequency and severity of sleep-related problems, subjective sleep quality, use of sleeping medications and daytime dysfunction. The seven component scores are summed to yield a total score with a range 0–21; higher scores indicate poorer sleep quality. Those scoring >5 are defined as poor sleepers. The clinical and psychometric properties of the measure have been assessed and it has been shown to have acceptable internal homogeneity and consistency in terms of test–retest reliability. It has also been validated by comparing good sleepers with patients with sleep or depressive disorders. Data from 52 healthy good sleepers in a study by Buysse et al. [14] was used as the normative comparison data in the current study.

To assess general well-being, *the short form SF-36* was used for all participants. This is a multipurpose, well validated, 36-item tool measuring generic functional health and quality of life, well-being and vitality. Eight subscales measure different aspects of health status: (1) limitations in physical activities because of health problems; (2) restrictions in usual role activities due to physical problems; (3) bodily pain, frequency and extent of interference; (4) vitality, measuring energy and fatigue; (5) general mental health (psychological distress and well-being); (6) limitations in social activities due to physical or emotional problems; (7) limitations in usual role activities due to emotional health; and (8) general health perceptions, including a question rating the amount of change in general health in a one-year period and limitations in physical, social and role activities due to physical or emotional health problems. Each sub-score produces a standardised score 0–100; a lower score indicates poorer health or greater disability while a higher score indicates the optimum health state [15]. Figures from 2056 British young people and adults were used as the general population norm in the current study [16].

Psychological well-being of both patients and main carers was established with the *Hospital Anxiety and Depression Scale (HADS)* [17]. This well validated brief self-report questionnaire contains 14 items assessing anxiety and depression in a non-psychiatric population. Over two separate subscales, seven questions rate anxiety (HADS-A) and seven questions rate depression (HADS-D) with answers rated on a four-point scale (0–3) where 0 is 'not at all' to 3 'very often indeed', and summed to give the subscale scores. The scale has been validated with scores between 0–7 indicating no risk, 8–10 borderline and 11 or more high risk for anxiety or depressive disorders [17]. Normative data from a UK population of 1792 individuals was used as a comparison for patients and carers in the present study [18].

To assess impact of illness on family life main carers completed the modified *Family Burden Interview Schedule* [19], a 25-item questionnaire designed to assess six domains of burden – financial impact, effect on family routines, on leisure, interactions, on the physical and on the mental health of family members – as well as a global subjective measure of burden; it has been found useful to assess impact on the family of different paediatric disorders [20,21]. Family function was further evaluated by main carers completing the *Family Assessment Device (FAD)* [22]. This 60-item self-report questionnaire is designed to assess individual perceptions of several dimensions of family functioning: problem solving, communication, roles, affective responsiveness, and behaviour control. Three of the six scales were used in the current study: (1) communication: assessing the verbal exchange of instrumental (daily decisions) and affective (emotional) information within the family as well as the clarity of communication, i.e., whether it is clear or camouflaged, direct or indirect; (2) problem solving: assessing the ability to solve instrumental (e.g., daily decisions such as finances) and affective problems (feelings and emotional experiences) that maintain effective family functioning; and (3) general functioning which assesses overall family functioning. Four alternative responses rate how well each statement describes the family: 'strongly agree', 'agree', 'disagree', and 'strongly disagree'. Cut-off values for family dysfunction are documented as 2.2 for Problem Solving and Communication, and 2.0 for General Functioning, with scores above the cut-off indicating poorer functioning. Data from a study on 1394 British families was used as the general population standard [23].

2.3. Qualitative interviews

The qualitative arm of the study used semi-structured interviews to explore the experiences of ventilation treatment of parents/carers, siblings and patients. The interviews enquired about (i) commencement of the condition; (ii) impact of treatment on health, activities, emotions; (iii) support; and (iv) the use of equipment.

2.4. Data analysis

The quantitative data was analysed using SPSS (Version 15.0 for Windows). Frequency data was calculated and compared with the available general population data. Non-parametric correlation analysis was carried out to examine the associations between sleep quality and illness, well-being and family variables.

All semi-structured interviews were audio recorded, the interviews were examined, and a coding frame was constructed.

3. Results

Out of 20 eligible families approached, 10 (2 with under one year (7 and 9 months, respectively), 4 between 1 and 5 years, and 4 with over 5 years experience of ventilation) agreed to complete questionnaires and 3 agreed to be interviewed (2 with over and 1 with under 5 years ventilation experience). The mean age of the group at the time of starting ventilation was 15.4 ± 3.8 years (range 11–21 years).

3.1. Patients

Socio-demographic and illness data about patients and the main carers are given in Table 1. The mean age of patient participants – all males and wheelchair dependent – was 19.7 ± 4.6 years. Parents estimated their son's educational achievement as average or above average and half were in active education. Psychological