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ORIGINAL RESEARCH

Prevalence of Fatigue, Pain, and Affective Disorders in Adults With Duchenne Muscular Dystrophy and Their Associations With Quality of Life

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Abstract

Objectives: To assess the prevalence of fatigue, pain, anxiety, and depression in adults with Duchenne muscular dystrophy (DMD), and to analyze their relationship with health-related quality of life.

Design: Cross-sectional study.

Setting: Home of participants.

Participants: Adults (N=80) with DMD.

Interventions: Not applicable.

Main Outcome Measures: Fatigue was assessed with the Fatigue Severity Scale; pain with 1 item of the Medical Outcomes Study 36-Item Short-Form Health Survey and by interview; and anxiety and depression by using the Hospital Anxiety and Depression Scale. Health-related quality of life was assessed using the World Health Organization Quality of Life Scale—Brief Version. Associations between these conditions and quality of life were assessed by means of univariate and multivariate logistic regression analyses.

Results: Symptoms of fatigue (40.5%), pain (73.4%), anxiety (24%), and depression (19%) were frequently found. Individuals often had multiple conditions. Fatigue was related to overall quality of life and to the quality-of-life domains of physical health and environment; anxiety was related to the psychological domain.

Conclusions: Fatigue, pain, anxiety, and depression, potentially treatable symptoms, occur frequently in adults with DMD and significantly influence health-related quality of life.

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Duchenne muscular dystrophy (DMD) is an X-linked, recessive neuromuscular disease in which there is an absence of the protein dystrophin, which among other things functions as a stabilizer of muscle cells, leading to progressive loss of muscle strength. The disease leads to severe physical disabilities and heavy dependency on care. In the past, most patients died in adolescence or young adulthood as a result of respiratory failure.[1]

As a result of improved care, especially the introduction of home mechanical ventilation, survival has increased significantly in the past few decades.[2,3] The probability of reaching the age of 30 years has been found to have risen up to 85%.[4] With an incidence of around 1 in 3500 live male births,[1] there now is a considerable and relatively new group of adults with DMD. Without exception, individuals in this group are severely disabled.

From studies of other neuromuscular diseases, it is known that patients experience a multitude of physical and psychological disorders. In a systematic review[5] of quality of life in adult neuromuscular disease, there was a high level of evidence that quality of life in adults with various muscle diseases was significantly affected by disease severity, pain, fatigue, and mood.

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In neuromuscular disorders, fatigue—described as an overwhelming sense of tiredness, lack of energy, and feeling of exhaustion—is a common symptom.[6-9] It is related in daily life to impairment caused by facioscapulohumeral muscular dystrophy, myotonic dystrophy, and hereditary motor and sensory neuropathy type 1.[10]

Pain, too, is known to be a significant problem in many chronic neuromuscular diseases.[11-13] Several studies[12,14] have found that pain has a negative effect on health-related quality of life in different neuromuscular diseases.

As for affective disorders, their prevalence differs among neuromuscular diagnoses[15]; depression and anxiety are moderately correlated to quality of life in adult patients with neuromuscular disease.[16] Depression appears to go underdiagnosed in patients with chronic somatic diseases.[17] In amyotrophic lateral sclerosis, depression plays a major role in determining quality of life.[18]

Identifying fatigue, pain, and affective disorders, when present, may allow effective treatment strategies to be used by health care providers. It is uncertain whether these conditions are more prevalent in adults with DMD than in the general population, how frequently they occur concurrently, and whether their presence is associated with lower health-related quality of life. In the present study, we posed the following 2 research questions: (1) What is the prevalence of fatigue, pain, and affective disorders in the adult DMD population? and (2) Are fatigue, pain, and affective disorders associated with lower health-related quality of life in this population?

Methods

Participant selection and procedure

This study was part of a larger cross-sectional study into the functioning and quality of life of adults with DMD and their informal caregivers.[19,20] Patients were recruited by all 4 Centres for Home Ventilation in the Netherlands, and by Dutch rehabilitation centers and the Dutch patient organization for neuromuscular diseases, Spierziekten Nederland. Inclusion criteria were a diagnosis of DMD and an age ≥20 years. Subjects were interviewed during 2 consecutive visits to their homes. The study was approved by the Medical Ethical Committee of the Erasmus University Medical Center, Rotterdam, the Netherlands. All subjects gave written informed consent for participation.

Measurements

We recorded age, data on physical situation (ventilation, gastrostomy, ambulation, hand function), and on participation (living situation, relational status, education level, work) to describe our population.

Fatigue was assessed using the Dutch version of the Fatigue Severity Scale. This is a self-rated questionnaire consisting of 9 statements on the effect of fatigue; for instance, "My motivation is influenced by fatigue," or "Fatigue interferes with my physical functioning." Mean scores on the 9 questions range from 1 (no signs of fatigue) to 7 (most disabling fatigue). The Fatigue Severity Scale has good validity and reliability.[21,22] A score ≥4 is considered indicative of significant fatigue.[23] We defined the presence of fatigue as a score ≥4; scores between 4 and 5 were defined as intermediate, and scores ≥5 as severe fatigue.[24]

Pain was assessed using the single-item ratings of pain of the Medical Outcomes Study 36-Item Short-Form Health Survey,[25] which asks about the presence of bodily pain in the last 4 weeks, with scores from 1 (no pain) to 6 (very serious pain). Scores ≥2 are considered to be indicative of the presence of pain[26]; scores of 2 to 4 are labeled intermediate, and scores of 5 and 6 severe. We also assessed the pain locations and the number of locations per individual, and asked how long pain had been present. We defined pain with a duration ≥3 months as chronic pain.

Affective disorders were assessed using the Hospital Anxiety and Depression Scale (HADS). The HADS, with separate subscales for anxiety (HADS-A) and depression (HADS-D), performs well in assessing symptom severity. Subjects are asked to score statements such as "I can sit at ease and feel relaxed" (anxiety) or "I feel cheerful" (depression), on a 4-point scale. On both subscales, scores can range from 0 to 21; scores of 8 to 11 indicate intermediate disorders, and ≥12 indicate clinical disorders.[27] As with pain and fatigue, we considered intermediate and clinical scores on the HADS subscales to be indicative of the presence of disorders. The HADS has been shown to be reliable and valid for assessing anxiety and depression.[28] In this study, both anxiety and depression scores ≥8 were considered to be indicative of the presence of disorders.

Quality of life was assessed using the World Health Organization Quality of Life Scale—Brief Version (WHOQOL-BREF). This is a questionnaire developed for cross-cultural comparison of quality of life relevant to global well-being.[29] It consists of 26 items, with 1 item measuring overall quality of life, 1 measuring satisfaction with health, and 24 items that can be combined into subscales for the 4 domains of physical functioning, psychological functioning, social relationships, and environment. All items are assessed with a 5-point Likert scale. Item scores are converted to domain scores ranging from 4 to 20, with higher scores representing better quality of life. The WHOQOL-BREF has good validity, internal consistency, and test-retest reliability.[30] In this study, the results on item 2 (satisfaction with health) were not used.

Statistics

Data were analyzed using SPSS version 20.[a] Descriptive statistics were used to assess the results for the various measurements. Correlations between fatigue, pain, anxiety, and depression were assessed using Spearman rho. Because the results on the WHOQOL-BREF had no normal distribution, we dichotomized the scores to compare persons in the lowest range with the others.[26] Overall quality of life was scored as good (item scores 4–5) versus poor (scores 1–3). For the WHOQOL-BREF domains, we used a cutoff of 1 SD below the reference mean. Subsequently, univariate and multivariate logistic regression analyses were performed with fatigue, pain, anxiety, and depression as independent variables. Since we found no significant associations of age, level of education, and ventilation type with the various domains of quality of life (results not shown), we did not adjust for these factors in the main analyses. In addition, we analyzed the difference in prevalence of fatigue, pain, anxiety, and depression between subgroups of men with good (WHOQOL-BREF item 1, score 4–5) and poor

List of abbreviations:

DMD	**Duchenne muscular dystrophy**
HADS	**Hospital Anxiety and Depression Scale**
WHOQOL-BREF	**World Health Organization Quality of Life Scale—Brief Version**