

Is functional dependence of Duchenne muscular dystrophy patients determinant of the quality of life and burden of their caregivers?

A dependência funcional de pacientes com distrofia muscular de Duchenne é fator determinante na qualidade de vida e na sobrecarga de seus cuidadores?

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ABSTRACT

Objective: The relationship between functional dependence and quality of life (QOL) in Duchenne muscular dystrophy (DMD) patients and burden and QOL in caregivers is not clear. This study investigated possible relationships between functional dependence/QOL of DMD patients and QOL/burden of caregivers. **Method:** This study included 35 boys (6-17 years) and respective caregivers (above 21 years). Caregivers answered to World Health Organization Quality of Life and Zarit Burden Interview questionnaires. Patients were assessed with the Motor Function Measure and the Autoquestionnaire *Qualité de vie Enfant Imagé*. Spearman correlations and linear regressions were run to investigate relationships between the variables. **Results:** The occurrence of lower QOL and higher burden among the caregivers of patients with Duchenne muscular dystrophy was evidenced. The functional dependence of patients was not considered a determinant factor. Higher caregivers' burden was related to lower caregivers' QOL and to higher patients' ages.

Keywords: Duchenne muscular dystrophy, behavior, quality of life, motor activity.

RESUMO

Objetivo: A relação entre qualidade de vida e função motora de pacientes com distrofia muscular de Duchenne (DMD) e sobrecarga e qualidade de vida (QV) dos cuidadores não está clara na literatura. Esse estudo investigou possíveis relações entre dependência funcional/QV de pacientes e sobrecarga/QV dos cuidadores em 35 meninos (6-17 anos) com DMD e respectivos cuidadores (acima de 21 anos). **Método:** Cada cuidador respondeu ao questionário de QV da Organização Mundial de Saúde e de sobrecarga de Zarit e o paciente foi avaliado com a medida de função motora e o *Autoquestionnaire Qualité de vie Enfant Imagé*. Correlações de Spearman e regressões lineares investigaram possíveis relações entre as variáveis. **Resultados:** A ocorrência de menor QV e maior sobrecarga entre cuidadores foi evidenciada, enquanto a dependência funcional dos pacientes não foi considerada fator determinante. **Conclusão:** Cuidadores com maior sobrecarga apresentaram menor QV relacionada à maior idade dos pacientes.

Palavras-chave: distrofia muscular de Duchenne, comportamento, qualidade de vida, atividade motora.

Duchenne muscular dystrophy (DMD) is a genetic X-linked recessive disease. Considering all the hereditary diseases, DMD has the second higher incidence¹, which varies from 1:3800 to 1:6200 among boys born alive². The absence of dystrophin results in a progressive degeneration of muscle fibers, causing muscle weakness. Gait loss and functional dependence occur in the second decade of life³. Although the

advances on treatment have increased the survival time, the functional independence is still poor, influencing biopsychosocial aspects of these children⁴ and their caregivers⁵.

The quality of life (QOL) of children with DMD is still controversial. Some studies have shown that it is reduced among children with DMD^{6,7}. Others have found no differences between the QOL of children with DMD and healthy

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children^{4,8,9}. However, most authors agree that the QOL does not change as the child grows older and gets more dependent^{7,9}, except Davis et al., who associated the reduction of the QOL of children and caregivers to the wheelchair dependence¹⁰. Opposite, the non-invasive ventilation dependence has not been related to the reduction of QOL in children⁴. The scores of QOL questionnaires in children are usually proportionally equivalent to the scores of their caregivers^{8,11}.

The main caregiver (in Brazil, usually the mother) shows higher scores in burden scales¹². Burden may impact on physical and mental health and influence the QOL. Higher anxiety, depression and stress have been described in parents of children with DMD^{13,14} influencing QOL⁵. Differently from the QOL, which does not alter, lower burden scores among caregivers are related to having ambulatory children (opposite to having wheelchair dependent children) and having a higher social support⁵. Parents of children with cognitive and/or behavioral impairments tend to have higher stress levels¹³.

The relationship between functional dependence of children with DMD and psychosocial aspects, such as the QOL of children and respective caregivers and the burden of caregivers is not clear in the literature. This study aimed to investigate these possible relationships between functional dependence of children with DMD, QOL of children with DMD and caregivers and burden of caregivers.

METHOD

Participants

This study included 35 boys (6-17 years) with molecular diagnosis of DMD and respective caregivers, recruited on the Neurology Division of the Clinics Hospital of the Faculty of Medicine of University of Sao Paulo. Data from one boy and his mother were excluded because she could not answer the World Health Organization Quality of Life Organization (WHOQOL-brief) questionnaire for being illiterate. Three caregivers had two children each, diagnosed with DMD. Therefore, the participants were 34 children and 31 caregivers. This study was approved by the local Ethics Committee. All participants formally consented to participate in the study.

Inclusion criteria comprehended being able to read and write (by self-report) and being able to communicate orally by writing. Among children, we included children aged between 6 and 17 years and among caregivers, above 21 years.

Procedure

Each participant was assessed individually in rooms containing a mat, a stretcher and the material required to answer the scales and questionnaires. Each caregiver

answered to the QOL and burden questionnaires while the child was assessed with the Motor Function Measure (MFM) and answered a QOL questionnaire.

Measures

Motor function

Children were assessed with the Brazilian version of the MFM^{15,16}. This scale includes 32 items, divided in three domains: standing and transferring; axial and proximal motor function; and distal motor function. Each item can be scored from 0 to 3 (0 is the worst and 3 the best possible score).

Quality of life of children

The QOL was measured by the *Autoquestionnaire Qualité de vie Enfant Imagé* (AUQEI). This questionnaire was elaborated by Manificant and Dazord¹⁷ and validated in Brazil by Assumpção et al.¹⁸. It allows the self-evaluation of QOL using four images of faces (very unhappy, unhappy, happy, very happy). It has 26 questions and the child is asked to say how he feels about daily life situations. The questions include family, social, activity, body functions and health relations. Each question is scored from 0 (very unhappy) to 3 (very happy). The maximum possible score is 78. The cutoff score is 48. Below this score, the QOL is considered reduced¹⁸.

Quality of life of caregivers

The QOL of the caregiver was assessed by the Brazilian brief version of the WHOQOL-brief^{9,20}. It consists of 26 questions, divided in 4 domains: physical, psychological, social and environmental. The physical domain involves questions about pain, discomfort and vitality. The psychological domain involves positive and negative aspects about learning, memory, attention, self-esteem, body image and appearance satisfaction. The third domain aims to identify social relations and the support network. The fourth domain involves environment, physical security, financial resources, leisure and transportation¹⁹. Each question has 5 possibilities of answer, varying from 1 to 5. Lower scores mean lower QOL and higher scores mean higher QOL. On question 26 (the last question), this relation is inverse. To our knowledge, there is no cutoff value for this questionnaire.

Burden of caregivers

The Brazilian version of the Zarit Burden Interview (ZBI) was used to evaluate the burden of the caregiver^{21,22}. It was originally developed to assess the caregivers of dependent older adults, but recent studies have used the questionnaire in children and verified good responsiveness and usability^{23,24}. The assessment consists of 4 factors: the impact of caregiving, interpersonal relations, caregiving expectancies and self-efficacy perception. It has 22 questions and each

question is scored by the caregiver from 0 to 4. The total score varies from 0 to 88²⁴. Higher scores indicate higher burden perception and some authors consider that scores above 21 indicate burden²⁵. The questions involve health, social life, personal life, financial situation, and emotions relation with the cared.

Statistical analysis

Statistical analysis was run using the software Statistica, version 12.0. Descriptive analysis included mean, standard deviation, median and P25-P75 of the caregiver QOL (WHOQOL-brief), caregiver burden, child's age and motor function and QOL. Scores correlations were tested using Spearman tests (one-tailed), because most variables were ordinal and had non-normal distribution. Correlations higher than 0.70 were considered strong, between 0.30 and 0.69 were considered moderate and below 0.29 were considered weak (significance level: $p < 0.05$).

A linear regression considered the WHOQOL as dependent variable, and ZBI, AUQEI, children's age, MFM, and caregiver's age, in that order, as independent variables. It aimed to verify which factors would explain the caregiver's QOL. Another regression considered ZBI as dependent variable and children's age, MFM, caregiver's age, and AUQEI, in that order, as independent variables. It aimed to verify which factors would explain the caregiver's burden. Durbin-Watson residuals and colinearity diagnosis were also considered to test the quality of the model.

RESULTS

Caregivers were 24 mothers (3 had 2 children), 5 fathers, 1 grandmother and 1 sister (mean age: 38 years). The children's mean age was 11 ± 3 years. None of the children was using non-invasive ventilation on time of the assessment. Seventeen boys (50%) were wheelchair dependent (mean age: 13 years) and the mean age of ambulatory children was 9 years. General sample characteristics are displayed on Table 1.

The QOL and burden of the caregivers were strongly correlated (Figure 1) showing a decrease on QOL for the ones with the higher burden perception. Fifty six percent of the caregivers can be considered burdened according the ZBI. The burden of the caregiver and the children's age were negatively correlated, (Figure 2) suggesting a higher burden in individuals with more years spent taking care of the child. Caregiver's burden was also correlated to children's QOL. Only twenty-three percent of the children (eight children) had low QOL in accordance with the cutoff value of AUQEI. Age was negatively correlated to QOL of the children and, as expected, motor function, in a way that younger children had better motor function and higher QOL. The

Table 1. Sample characteristics of children with DMD and caregivers (n = 34).

Variable	N
Children's age, mean (min/max)	11 (6-17)
Caregiver's age, mean (min/max)	38 (20-52)
Mothers interviewed, n (%)	24 (71)
Locomotion, yes (%)	17 (50)
MFM score, mean (SD)	64.0 (21.3)
AUQEI score, mean (SD)	51.8 (6.7)
WHOQOL score, mean (SD)	14.0 (2.0)
ZBI score, mean (SD)	26.3 (10.1)

DMD Duchenne muscular dystrophy; MFM: Motor function measure – total score; AUQEI: *Autoquestionnaire Qualité de vie Enfant Imagé*; WHOQOL: World Health Organization Quality of Life Assessment – brief version; ZBI: Zarit Burden Interview; SD: Standard deviation; min: Minimal; max: Maximum; n: Number.

children's QOL was also correlated to the caregiver's age. Table 2 shows all these correlations and Spearman coefficients ($p < 0.05$).

The linear regression analysis between the QOL and burden of the caregiver (ZBI) and the QOL of the child evidenced that the QOL of the child was not relevant for the model. Therefore, this variable was excluded in this analysis. The burden of the caregiver explained 69% of the QOL of the caregiver variability (R square change = 0.477). Considering ZBI and patient's age, this variable was not relevant and was excluded. When the motor functional independence was introduced in the analysis, it explained 28% of the QOL, and remained as an important factor when the mother's age was included, being this last factor not relevant to the model. All quality indexes were

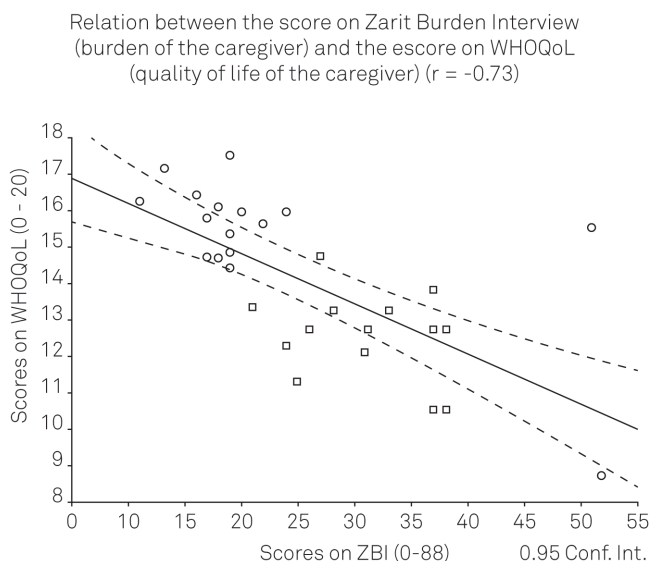


Figure 1. Relation between the score on Zarit Burden Interview (burden of the caregiver) and the score on WHOQoL (quality of life of the caregiver) ($r = -0.73$).

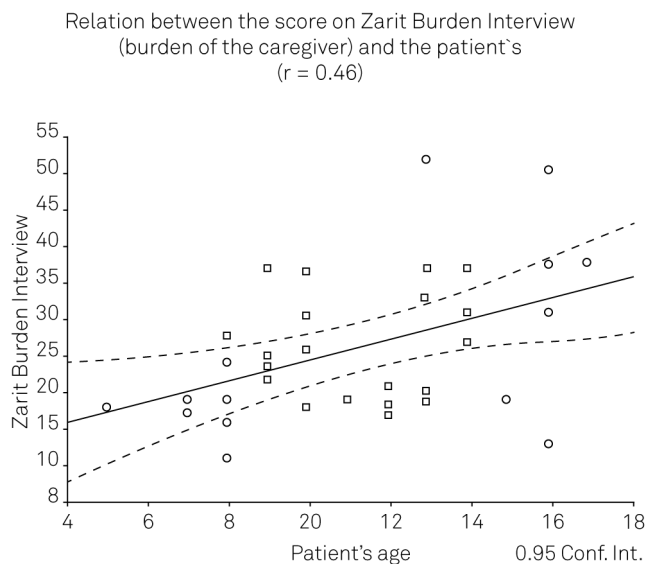


Figure 2. Relation between the score on Zarit Burden Interview (burden of the caregiver) and patient's age ($r = -0.46$).

suitable for the model (Durbin-Watson = 1.59 and colinearity tolerance = 1.00).

The linear regression analysis considering the burden of the caregiver, the age and the motor function of the children evidenced that the motor function of the child is not relevant to the model. The age explained 45% of the burden referred by the caregivers (R square change = 1.97), and all the other factors were not relevant for model. All quality indexes of the model were suitable (Durbin-Watson = 2.14 and colinearity tolerance = 1.00).

DISCUSSION

The present study demonstrated that the QOL of the caregivers of children with DMD is related mostly to their burden. This relation has been previously described in studies with the primary caregivers of adults and older adults^{26,27}, but no previous study investigated this relation in people responsible for children with neuromuscular diseases.

Our results showed that the burden of the caregivers is mostly explained by the children's age in a way that caregivers of older children showed more burden than the ones of younger children. On the other hand, the lower functional independence of older boys did not show correlation to the burden, what make us believe that the time taking care of these patients burdens the caregivers and decreases their QOL. Thompson et al. did not find any interference of the age of children with DMD on the caregivers stress and anxiety¹⁴. However, that study included younger boys (7-12 years) and the present study included a broader age range, with a higher median in our sample (6-17 years). Therefore, the contribution of the present study is to show a higher influence of the time caring for the child and the burden of the caregivers. We could also demonstrate that the burden is associated to the QOL reduction of both caregivers and children with DMD.

Few studies have investigated the burden of caregivers of children with DMD. No previous study has associated the QOL to the burden, nor to characteristics of the children. We observed that 69% of the QOL of the caregivers is explained by the burden of taking care of children with DMD. Kenneson and Bobo observed high scores in questionnaires investigating stress and anxiety among caregivers of children with DMD and Becker muscular dystrophy. In that study, almost 40% of the caregivers reported being unsatisfied with their lives and 50% showed a high burden. However, the authors concluded that only socioeconomical aspects were associated to the burden⁵. Stress, depression and anxiety have been described in groups of caregivers of children with DMD, and have been related to the introspective behavior of these children^{13,14}.

In a study with adults with DMD, a substantial burden was observed in caregivers. This burden of the caregivers was associated to being responsible for patients with tracheostomy and needing ventilation. Also, patients whose caregivers scored higher in anxiety questionnaires also showed higher anxiety scores²⁸. In the present study, none of the patients were in use of mechanical ventilation. This factor is associated with the low social condition of our sample, and the difficulty in acquiring the ventilator by public

Table 2. Spearman correlations between the caregiver's QOL, age, motor function and children's QOL in relation to the caregivers' burden and to each other (n = 34).

	WHOQOL	ZBI	MFM	AUQEI	Patient's age	Caregiver's age
WHOQOL	-	-0.729*	-0.116	0.234	-0.133	-0.008
ZBI		-	-0.235	-0.308*	0.459*	0.092
MFM			-	0.276	-0.672*	-0.123
AUQEI				-	-0.292	-0.447*
Patient's age					-	0.197
Caregiver's age						-

*p < 0.05. QOL: Quality of life; MFM: Motor function measure – total score; AUQEI: *Autoquestionnaire Qualité de vie Enfant Imagé*; WHOQOL: World Health Organization Quality of Life Assessment – brief version; ZBI: Zarit Burden Interview.

resources in our country. Boyer et al. verified that there is a relation between the burden of the caregivers of patients with many kinds of muscular dystrophy and the social aspects of the QOL questionnaire. They also observed that caregivers younger than 48 years showed higher burden levels²⁹. In the present study, the mean age of the caregivers was 38 years old, which agrees with the results of Boyer et al., whose study described a mean age of 32.7 years. On the other hand, this factor was not relevant neither for the caregiver's QOL nor their burden. We believe that this opposite result may be due to the high impact of the children's age over the caregiver's burden, which made the caregiver's age become a factor not relevant for the regression model.

In another limiting neurological disease, the cerebral palsy, this influence of functional aspects of the child on the caregivers QOL has been previously investigated. The functional limitations of the children, measured by a functional scale (the Gross Motor Function Classification Scale) did not interfere on the QOL of the caregivers^{30,31}. We found the same results among children with DMD and caregivers. Nereo et al. verified that the high stress level of parents of children with DMD is determined mainly by the

family behavioral and social aspects¹³. In the present study we verified that the motor and functional limitations were not the factors with higher impact on the caregivers' burden and QOL, nor they determined a lower QOL of the child. Previous studies had shown high QOL of children, with no relation with the disease progression⁴. In the present study we observed that the QOL of patients with DMD was only related to caregiver's age in a way that higher QOL was observed in boys who were cared by younger caregivers.

To sum up, the present study evidences the occurrence of lower QOL and higher burden in caregivers of patients with DMD. It also clarifies that the origin of the burden is related specifically to the time caring for the children, and it is not correlated to the loss of functional independence of the patients. This information should be considered in order to benefit the relations of families with children with DMD. An intervention based on the whole family and not only on the child with DMD should be considered³². Besides, when the child is diagnosed with DMD, the parents should be oriented to share the responsibilities of caring for the child, in order to increase their QOL and reduce their burden.

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