Acta Myologica • 2014; XXXIII: p. 136-143

Psychological and practical difficulties among parents and healthy siblings of children with Duchenne vs. Becker muscular dystrophy: an Italian comparative study

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This study explored the burden in parents and healthy siblings of 4-17 year-old patients with Duchenne (DMD) and Becker (BMD) muscular dystrophies, and whether the burden varied according to clinical aspects and social resources.

Data on socio-demographic characteristics, patient's clinical history, parent and healthy children burden, and on parent's social resources were collected using self-reported questionnaires administered to 336 parents of patients with DMD (246) and BMD (90).

Parents of patients with DMD reported higher burden than those of patients with BMD, especially concerning feeling of loss (84.3% DMD vs. 57.4% BMD), stigma (44.2% DMD vs. 5.5% BMD) and neglect of hobbies (69.0% DMD vs. 32.5% BMD). Despite the burden, 66% DMD and 62.4% BMD parents stated the caregiving experience had a positive impact on their lives. A minority of parents believed MD has a negative influence on the psychological well-being (31.0% DMD vs. 12.8% BMD), and social life of unaffected children (25.7% vs. 18.4%).

In the DMD group, burden correlated with duration of illness and parent age, and burden was higher among parents with lower social contacts and support in emergencies. In DMD, difficulties among healthy children were reported as higher by parents who were older, had higher burden and lower social contacts. In both groups, burden increased in relation to patient disability. These findings underline that the psychological support to be provided to parents of patients with MD, should take into account clinical features of the disease.

Key words: Duchenne muscular dystrophy, Becker muscular dystrophy, parents, healthy siblings, burden, social network

Introduction

Muscular Dystrophies (MDs) are degenerative, rare muscle diseases leading to progressive restriction of functional autonomy (1). Although curative therapy is not yet available, the improvement of standard care has led to a considerable increase in patients' life expectancy (2).

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Duchenne Muscular Dystrophy (DMD) – the most severe form of MD – is due to X-linked dystrophin gene mutations and affects about one in 5.000 males (1). Typically, symptoms of DMD manifest between 2 and 5 years of life, ambulation is lost by 12 years, and death mostly occurs in the second or third decade of life (1). Becker MD (BMD), the allelic milder form, affects about one in 20.000 males. In BMD, muscle symptoms usually onset in the second decade, walking autonomy is preserved up to the fifth or sixth decade, and life expectancy is not significantly reduced, unless cardiomyopathy occurs (3).

Most patients with MD, even those affected by severe forms, live at home and receive daily assistance from their relatives. Home care facilitates patients' maintenance of an acceptable daily routine for as long as possible, while caregivers receive a multifaceted experience (4-6).

Consequences of family caregiving in chronic diseases are commonly named "family burden" and subdivided into "practical" and "psychological" burden (7). Practical burden refers to problems such as disruption of family relationships, constraints in social, leisure, and work activities, and financial difficulties. Psychological burden describes the reactions that family members experience, e.g. feeling of loss, sadness, tension, and feeling unable to cope with the situation. Family burden has been scarcely explored in MDs, differently from cancer (8), dementia (9) and mental disorders (10). Available data reveal that caregivers of patients with MDs may perceive moderate to high levels of stress, and have frequent feelings of guilt, sadness and depression related to the patient's condition (6, 11, 12). Moreover, the caregivers frequently face financial difficulties due to costs of care and constraints of work activities, neglect other family members and reduce their own social activities (4, 6, 7, 12-14). Mothers, low-income families, unemployed relatives, and relatives of patients with high disability and severe MD present higher levels of burden (6). Conversely, relatives who have adequate coping skills, high self-esteem, and a supportive social network perceive lower burden and identify more valuable benefits from the caregiving experience (12, 14). These findings can be interpreted within the framework of Lazarus and Folkman's transactional model (15), which postulates that an individual's adaptation to an event is a process based on primary and secondary cognitive appraisal. In regards to MD, primary appraisal has to do with the perception of the stress and consequences associated with MD, while secondary appraisal implies the development of strategies to cope with the difficulties of caring. In this model, adaptation is significantly influenced by internal factors (i.e.key-relative's attitude toward the patient) and external resources (i.e. availability of social and professional support) (14).

Home care for a child with MD involves the whole family, including minor unaffected siblings. However, little information on the psychological adjustment and the practical consequences of family care for healthy children is available. Studies on unaffected siblings of children with other severe pediatric illnesses suggest that emotional distress and behavioral problems may be significantly high in healthy siblings, and in part related to adult relatives' burden (16-18). In a study addressing psychological adjustment in 46 minor siblings of DMD children (19), 52% and 44% of siblings reported "great" or "some" deal of involvement in their brother's care, respectively, while 37% and 35% stated they missed special activities and/or daily activities due to a patient's care. Furthermore, a higher proportion of healthy siblings reached the high-risk threshold for emotional problems, and – as rated by parents – more than twice (19%) the high-risk cut-off for a psychiatric disorder, compared to the normative sample. Psychological symptoms were found to be weakly/moderately related to a patient's age and closeness in age to the affected sibling, adult relatives' burden, and family communication skills. However, DMD was also found to be associated with positive psychological adjustment in the family (20).

Research on MDs burden has several weaknesses. The few available studies have only investigated family burden in DMD (5, 12) and were carried out in North-America (6-13), limiting the generalization of the results in contexts with different health care policies (21). The poor data have probably slowed down the development and dissemination of targeted interventions to support patients and families in the routine, as the allocation of economic and staff resources.

In 2012, within the framework of the Telethon-UILDM Italian National Program for Clinical Research in Muscular Diseases, we performed a national survey on the condition of families of patients with DMD, BMD, or Limb-Girdle MD (LGMDs). Data on 502 families of patients aged 4 to 25, revealed that feelings of loss and sadness were present in 77% and 74 % of relatives, respectively, while constraints to leisure activities were present in 59%. Burden was higher among relatives of patients with lower functional abilities, who were older in age, and suffering from DMD, and among those who were more involved in a patient's daily care or who perceive lower social support. Psychological benefits were acknowledged by 88% of the relatives, particularly those who perceived a higher level of professional and social support (7, 14).

Based on the national data bank mentioned above, this paper is focused on the difficulties experienced by 246 parents of minors with DMD and 90 parents of minors with BMD, and the parents' perception of difficulties in minor unaffected children. In particular, the study

aims to verify whether: the burden is higher in relatives of patients with DMD than in those of patients with BMD, even in early and intermediate stages of the diseases; the difficulties experienced by the parents and by healthy siblings – as perceived by the parents – are higher among parents who are older and have lower social resources and with affected children having longer duration of illness, higher disability and older age.

Methods

Design of the study

The study was carried out in Italy in eight tertiary neuromuscular centers from January to December 2012. In each center, key-relatives (i.e., the relative spending more daily time in contact with the patient and being more involved in his/her care) of 4-25 year patients who had a diagnosis of DMD, BMD, or LGMD, and lived with at least one relative 18-80 years-old, were consecutively contacted and asked to participate. In occasion of the patient's clinical scheduled control, key-relatives were interviewed – after written informed consent – by a trained researcher on: a) main socio-demographic characteristics of the family and clinical history of the patient through an ad-hoc designed schedule; b) patient's functional abilities, according to Barthel Index (BI) (22); c) treatments and support received by the patient and his/her family through the Muscular Dystrophy Care Schedule (MD-CS). Furthermore, they were invited to fill in the Family Problems Questionnaire (FPQ) (21) and the Social Network Questionnaire (SNQ) (21). Among the 502 key-relatives who participated in the study, those having one 4 to 17 year-old child with DMD or BMD, were subsequently extrapolated for the aims of this paper.

The protocol of the study was approved by the Ethic Committee of the Second University of Naples (coordinating centre) and accepted by the Ethical Committee of each center.

Instruments description

BI assesses patient's global degree of independence in daily activities on a 0-100 score (from 0 "totally dependent" to 100 "totally independent"). In this study, the inter-rater reliability in BI scoring, measured by Cohen's kappa coefficient, ranged from 1 to 0.90 for 9 BI items and was equal to 0.67 for the lasting one. MD-CS collects information on pharmacological, socio-rehabilitative, and psychotherapeutic interventions received by the patient, and professional and welfare support provided to the family in the last six months. FPQ is a 34-item tool exploring: a-b) psychological and practical burden; c-d) social and professional support to families in patient's emergencies; and e) relative's positive attitude toward the patient. Two additional items explore respondent's perception of psychological and social consequences in minor children. SNQ is a 15-item tool exploring: a) quality and frequency of social contacts; b-c) practical and emotional support; and d) quality of an intimate relationship. FPQ and SNQ are self-reported and contain items rated on a 4-level scale from 1 "never" to 4 "always". Mean subscale scores, ranging from 1 to 4, are also computed. FPQ and SNQ have been initially developed for schizophrenia and validated in five languages (English, Italian, Portuguese, Greek, and German) within the framework of a EC study on schizophrenia (21) (Cohen's kappa coefficient: 0.50 to 1 in 79% of FPQ items, and in 69% of SNQ items; Crombach's alpha: 0.61 to 0.88 of FPQ subscales, and 0.56 to 0.75 of SNQ subscales; factor analysis: 45% explained variance in FPQ, and 56% in SNQ).The main psychometric properties of FPQ and SNQ were further explored in samples of relatives of patients with physical diseases (10) and found to be consistent with those of the schizophrenia group (FPQ subscales alpha ranging between 0.91 and 0.65; explained variance 74%; SNQ subscales alpha value ranging between 0.75 and 0.59; percentage of the explained variance 72%). In the whole GUP10002 study sample (7), Crombach's alpha on FPQ and SNQ subscales were found consistent with previous measurements (0.63 to 0.86 in FPQ subscales and 0.68 to 0.79 in SNQ subscales). In this paper, items from FPQ a-c subscale items (alpha from 0.66 to 0.87 in this sample) and additional items on siblings difficulties (alpha: 0.72), and SNQ a) subscale items (alpha: 0.69) have been reported. Furthermore, for this study, a burden total score was also computed (alpha: 0.86).

Statistical analysis

χ^2 and analysis of variance were used, as appropriate, to test differences in nominal and ordinal variables between DMD and BMD samples. χ^2 was also used to compare the two samples with regards to FPQ burden items. Analysis of variance was used to compare the two samples in their mean scores of parents' burden. In each group, correlations of parents' burden with perceived difficulties in unaffected siblings, were explored by Spearman's r correlation. The same test was used to explore the relationships of parents' burden and perceived difficulties in healthy children with parents' social contacts and social support in emergencies, parents' age, and patient's age, BI and duration of illness. Because of the large number of analyses, only results at the $p < 0.05$ with Bonferroni correction are reported, to reduce the probability of type I errors (false positives).

Results

Among the 246 children with DMD and 90 with BMD, the majority attended school and had healthy siblings (Table 1). Children with DMD were younger ($F = 13.9$, df 1, 334, $p < .05$), had lower levels of functional autonomy ($F = 95.7$, df 1, 334, $p < .05$) and more frequently received economic welfare benefits (177, 72.0% vs. 31, 34.4%, $\chi^2 = 39.3$, df 1, $p < .05$) than patients with BMD. Two-hundred-eight patients with DMD (84.6%) and 39 (43.3%) with BMD ($\chi^2 = 57.5$, df 1, $p < .05$) were in drug treatment (corticosteroids: 168 (68.3%) of DMD vs 7 (7.8%) of BMD; bone metabolism drugs: 92 (37.4%) vs. 7 (7.8%); cardiologic drugs: 80 (32.5%) vs. 22 (24.4%); gastric drugs: 48 (19.5%) vs. 4 (4.4%); neurological drugs: 5 (2.0%) vs. 1 (1.1%); pulmonary drugs: 4 (1.6%) vs. 1 (1.1%)) while 203 (82.5%) patients with DMD and 26 (28.9%) with BMD ($\chi^2 = 87.3$, df 1, $p < .05$) attended rehabilitation programs. Most participating parents were mothers, and had a middle to high educational level (Table 1). DMD parents spent more daily hours in patient caregiving than BMD parents ($F = 43.2$, 1, 334, $p < .05$). On average, burden was significantly higher among parents of children with DMD [1.8 (0.5), vs. 1.4 (0.4), $F=44.7$, df 1, 334, $p < .05$]. In particular, the feeling of loss was reported by 84.3% of parents in the DMD group vs. 57.4% of parents in the BMD group (Table 2). Perception of a stigma in a public setting was reported by 44.2% of DMD parents, while it was almost nonexistent in BMD group (5.5%). Moreover, 59.3% DMD vs. 30.3% BMD parents agreed with the statement "*I felt that I would not be able to bear the situation longer*" and 55% DMD vs. 29.2% BMD parents believed that if the patient was not sick, everything would be fine in their family. As far as practical consequences of caregiving (Table 3), differences between DMD and BMD group were particularly relevant in regard to the need to awaken during the night (47.3% vs. 17.7%), a neglect of hobbies (69.0% vs. 32.5%), difficulties in work/household activities (55.5% vs. 18.9%), taking holidays (38.9% vs. 12.0%), and financial difficulties (42.0% vs. 17.8%).

Forty-one (31.0%) DMD and 5 (12.8%) BMD parents believed that the patient's condition negatively influenced the psychological well-being of unaffected children, while 34 (25.7%) and 7 (18.4%) respectively felt a negative influence on the sibling's social life (Table 4).

Despite difficulties, most key relatives (66.0% DMD and 62.4% BMD parents) considered their caregiving experience to have a positive impact on their lives. Both DMD and BMD parents mentioned "*personal grown*" (73.6% vs. 63.1%, e.g., *'I learned that difficulties of life help you to grow")*, "*resilience*" (15.9% vs. 18.5%, e.g., *"I learned to have more strength to fight for the people I love*"), and "*altruism*" (15.9% vs. 15.4%, e.g., "*I get involved in helping people in a condition similar to mine"*) among the psychological benefits.

Furthermore, the majority of parents (70% DMD and 73.1% BMD) claimed to have at least two trustworthy

Table 1. Characteristics of patients with DMD and BMD and their parents.

	DMD (N = 246)	BMD (N = 90)
Patients		
Age, mean (SD) years	10.0 (3.7)	11.9 (3.6)
School attendance, N (%) yes	233 (94.7)	89 (98.9)
Minor healthy siblings, N (%) yes	134 (54.5)	45 (50.0)
Age of older healthy sibling, mean (SD) years	9.6 (4.4)	9.5 (4.7)
Duration of symptoms, mean (SD) years	6.8 (3.9)	7.5 (4.1)
BI, mean (SD)*	65.6 (28.0)	95.4 (11.3)
Parents		
Mothers, N (%)	205 (83.3)	78 (86.7)
Fathers, N (%)	41 (16.7)	12 (13.3)
Age, mean (SD) years	41.2 (6.2)	43.3 (6.6)
Marital status, cohabitant/spouse N (%)	217 (88.2)	80 (88.9)
Education, N (%) Primary school Secondary school High school University	 11 (4.5) 89 (36.2) 119 (48.4) 27 (10.9)	 3 (3.3) 37 (41.1) 39 (43.3) 11 (11.2)
Currently employed, N (%) yes	133 (54.1)	55 (61.1)
Daily hours in patient's caregiving , mean (SD)*	6.3 (4.1)	3.3 (2.7)

DMD = Duchenne Muscular Dystrophy; BMD = Becker Muscular Dystrophy; differences between the two groups explored by X or ANOVA test, * $p < .05$ with Bonferroni correction

Table 2. Psychological difficulties in DMD vs. BMD groups (N = 246 vs. N = 90).

Items – section a of FPQ	MD Type	Always N (%)	Often N (%)	SometimesN (%)	Never N (%)	Mean (SD)	χ2	MissingN
I felt that I would not be able to bear this situation much longer	DMD	8 (3.2)	35 (14.3)	103 (42.0)	99 (40.4)	1.8 (.0)		1
	BMD	2 (2.2)	4 (4.5)	21 (23.6)	62 (69.7)	1.4 (.7)	23.2*	1
I cried or felt depressed	DMD	7 (2.8)	59 (24.0)	130 (52.8)	50 (20.3)	2.1 (.7)		0
	BMD	4 (4.5)	8 (9.0)	49 (55.1)	28 (31.5)	1.9 (.7)	11.4	1
I worry for the future of other family members	DMD	21 (8.6)	41 (16.8)	119 (48.8)	63 (25.8)	2.1 (.9)		2
	BMD	3 (3.3)	11 (12.2)	45 (50.0)	31 (34.4)	1.8 (.8)	5.2	0
When I went to a public place with my ill relative, I felt that everyone was watching us	DMD	19 (7.9)	20 (8.3)	68 (28.1)	135 (55.8)	1.7 (.9)		4
	BMD	1 (1.1)	0	4 (4.4)	85 (94.4)	1.1 (.9)	44.1*	0
I feel guilty because I believe that I or my spouse may have passed on the illness to our relative	DMD	19 (7.8)	30 (12.2)	80 (32.7)	116 (47.3)	1.8 (.9)		1
	BMD	15 (17.0)	18 (20.5)	29 (33.0)	26 (29.5)	2.2 (1.1)	13.3	2
I think that if our relative didn't have this problem, everything would be all right in our family	DMD	39 (16.0)	32 (13.1)	64 (26.2)	109 (44.7)	2.0 (1.1)		2
	BMD	4 (4.5)	3 (5.6)	17 (19.1)	63 (70.8)	1.4 (.8)	19.9*	3
When I think of how our ill relative was beforehand and how he/she is now, I feel disappointed	DMD	75 (31.0)	50 (20.7)	79 (32.6)	38 (15.7)	2.7 (1.1)		3
	BMD	12 (13.8)	11 (12.6)	27 (31.0)	37 (42.5)	2.0 (1.0)	29.6*	3

DMD = Duchenne Muscular Dystrophy; BMD = Becker Muscular Dystrophy;*, p < .05 with Bonferroni correction

Table 3. Practical difficulties in DMD vs. BMD groups (N = 246 vs. N = 90).

Items – section b of FPQ	MD Type	Always N (%)	Often N (%)	SometimesN (%)	Never N (%)	Mean (SD)	χ2	MissingN
I have had to wake up during the night	DMD	29 (11.8)	20 (8.2)	67 (27.3)	129 (52.7)	1.8 (1.0)		1
	BMD	1 (1.1)	4 (4.4)	11 (12.2)	74 (82.2)	1.2 (.6)	25.7*	0
I have had to neglect my hobbies and things I like doing in my free time	DMD	41 (16.8)	40 (16.4)	87 (35.7)	76 (31.0)	2.2 (1.0)		2
	BMD	2 (2.2)	8 (9.0)	19 (21.3)	60 (67.4)	1.5 (.7)	38.4*	1
I have had difficulty in going on Sunday outings	DMD	16 (7.8)	21 (10.3)	43 (21.1)	124 (60.8)	1.6 (.9)		42
	BMD	0	3 (3.7)	11 (13.6)	67 (82.7)	1.2 (.5)	15.2	9
I found it difficult to have friends at home	DMD	2 (0.8)	11 (4.5)	26 (10.7)	205 (84.0)	1.2 (.5)		2
	BMD	1 (1.1)	0	2 (2.2)	87 (96.7)	1.1 (.3)	10.9	0
I found it difficult to meet friends and people I like to spend my leisure time	DMD	6 (2.5)	22 (9.0)	50 (20.5)	166 (68.0)	1.5 (.8)		2
	BMD	3 (3.4)	0	9 (10.1)	77 (86.5)	1.2 (.6)	15.2	1
I found it difficult to carry out my usual work or household activities	DMD	11 (4.5)	31 (12.7)	94 (38.4)	109 (44.5)	1.8 (.8)		1
	BMD	0	3 (3.3)	14 (15.6)	73 (81.1)	1.2 (.5)	36.5*	0
I had to neglect other family members	DMD	1 (0.4)	27 (11.5)	75 (31.9)	132 (56.2)	1.6 (.7)		11
	BMD	0	2 (2.2)	18 (20.2)	69 (77.5)	1.2 (.5)	14.3	1
I had difficulty in going on holiday	DMD	28 (13.8)	21 (10.3)	30 (14.8)	124 (61.1)	1.1 (.1)		43
	BMD	1 (1.2)	2 (2.4)	7 (8.4)	73 (88.0)	.5 (.0)	21.8*	7
I had economic difficulties	DMD	12 (4.9)	20 (8.2)	71 (29.0)	142 (58.0)	1.6 (.8)		1
	BMD	1 (1.1)	1 (1.1)	14 (15.6)	74 (82.2)	1.2 (.5)	18.3*	0

DMD = Duchenne Muscular Dystrophy; BMD = Becker Muscular Dystrophy; * p <. 05 with Bonferroni correction

Table 4. Difficulties in healthy siblings as perceived by parents (N = 134 DMD vs. N = 45 BMD).

Items – additional section of FPQ	MD Type	Always N (%)	Often N (%)	Sometimes N (%)	Never N (%)	Mean (SD)	$\chi 2$	Missing N
I feel that the presence of S affects negatively the psychological well-being of my children (e.g., I see them crying, being fearful, aggressive, shy)	DMD	2 (1.5)	6 (4.5)	33 (25.0)	91 (68.9)	1.4 (.6)		2
	BMD	1 (2.6)	0	4 (10.3)	34 (87.2)	1.2 (.5)	6.3	6
I feel that the presence of S affects negatively the social life of my children (school performance, leisure activities, etc.)	DMD	3 (2.3)	5 (3.8)	26 (19.7)	98 (74.2)	1.3 (.7)		2
	BMD	0	0	7 (18.4)	31 (81.6)	1.2 (.4)	2.5	7

DMD = Duchenne Muscular Dystrophy; BMD = Becker Muscular Dystrophy

friends, and considered two or more relatives as trustworthy friends (78.0% DMD and 78.9% BMD). Moreover, in case of patient's emergencies, parents stated to have at least two friends/relatives (63.4% DMD and 58.8% BMD), on which rely, and to be confident to be helped by them always or often (72.3% DMD and 80.0% BMD). Furthermore, in the last two months, 50.0% of DMD relatives and 55.6% of BMD had been in contact with friends face to face or by phone, most days.

Burden was higher among parents of patients with lower functional autonomy (DMD burden total score: $r = -.50$, BMD burden total score: $r = -.38$, $p < .05$), in both groups. In the DMD group, burden correlated with duration of illness ($r = .32$, $p < .05$), patient's age ($r = .36$, $p < .05$) and parents' age ($r = .30$, $p < .05$). Furthermore, burden was higher among parents with fewer social contacts ($r = -.28$), and lower social support in emergencies ($r = -.51$, $p < .05$). In the same group, difficulties in healthy siblings were higher among children whose parents were older ($r = .33$, $p < .05$) and with fewer social contacts ($r = -.27$, $p < .05$).

Discussion

The results of this study confirm that parents of children with DMD experience higher difficulties than those of children with BMD, even when the patient's functional ability is still relatively preserved. The main object of concern – significantly higher in DMD vs. BMD group – is the frequent feeling of loss and being inadequate to bear the situation and the conviction that whole family is influenced by the patient's condition. Forty-four percent of DMD parents felt to be observed in public places when they are with the sick child, but this feeling is virtually absent in the BMD group. Perceived stigma, a phenomenon largely investigated in mental illness (23-25) and rarely considered in physical illness (26), may negatively influence parents' and patient's quality of life over time. In particular, the stigma may lead to family social withdrawal (23), and may be associated with feelings of depression and guilt among parents (23). Moreover, the parents' perception of a stigma may contribute to a reduction of social contacts in patients, and negatively influence the adherence to treatment (25).

Differences in the onset and clinical course of DMD vs. BMD can explain why a lower parental burden was observed in the BMD group. In DMD, the early onset of symptoms may influence the mother-child relationship, and may become an obstacle to the child's social experiences (26, 27). Conversely, in BMD the later onset has a limited interference with patient's emotional development in childhood and adolescence, and the slow progression allows a gradual adaptation of parents and patients to the disability itself (27).

Furthermore, a clear relationship between increased levels of burden and reduction in parents' social ties was found only in the DMD group, though the social network did not differ between the two groups. It is likely that, as DMD progresses, parents feel overwhelmed by their caregiving role, and too exhausted to be involved in social activities. This situation may lead to a vicious cycle in which a progressive reduction of social network exposes the parents to greater levels of burden over time, with consequent further social withdrawal (10).

While parental burden is higher in the DMD, the difficulties observed by parents in their minor healthy children are similar in the two groups and relatively mild. It is likely that parents tend to protect unaffected children, not involving them in the care of the patient as longer as possible (19). However, when the disease progresses and burden increases (16), even healthy siblings are invited to take care of the patient, and this may lead to the onset of practical and psychological difficulties (20).

This study also showed that about two-thirds of parents, both DMD and BMD, acknowledged psychological benefits in their caregiving experience, especially "personal growth" and an increased sense of strength against adversity. It's likely that – as postulated by Lazarus and Folkman's model (15) – when relatives feel they can manage the practical difficulties of care, they do not overcome the individual threshold of stress tolerance, and may also consider the positive aspects of caregiving.

This study presents some limitations: (a) the assessment of burden only in the key-parent – mainly the mother, as is customary in Italy – does not allow us to estimate the influence of the caregiving on the parental couple (5, 28); (b) the study did not assess psychological adjustment in healthy siblings themselves (19); c) the lack of data from normal and healthy population collected by FPQ and SNQ; and d) the cross-sectional design of the study that does not allow inferences regarding the evolution of burden or whether external resources may influence the burden or vice versa. These limitations will be addressed in future studies, using an online assessment to overcome the logistical difficulties found in the involvement of more relatives per patient.

On the other hand, the main strengths of this comparative study exist in the large sample size, the participation of centers located in different geographical areas of Italy, and the use of validated assessment tools (21, 22).

The results of this study underline the need to differentiate the type of parental support, taking into account the clinical features of MDs. In the case of BMD, education on the illness and its course could be sufficient to facilitate a parent's adaptation and their active involvement in care (29). Conversely, in the more severe form of MD, targeted psychological support should be provided to parents in the different stages of the disorder, according to family need (29). Physicians, in collaboration with psychologists, should be trained in addressing parents' psychological reactions to diagnosis (30) and disease course. Furthermore, these professionals should maintain a hope-oriented approach to provide parents with education on MD, helping them to see the child "beyond the illness" and to communicate with unaffected children about the diagnosis (14, 19, 20, 31). Finally, they should prompt parents to, as useful coping strategies, adopt a problem-solving approach to deal with difficulties, to carve out time for their social contacts and to joint associations (32).

In conclusion, our results highlight that parents' and healthy siblings reactions to MDs vary in relation to type of the disease and parents' social resources. Moreover, the study focuses on aspects that are usually neglected by physicians and that would require planned professional training and appropriate resource allocation (33).

Acknowledgments

This study was supported by a Telethon UILDM grant (Grant n. GUP10002). We are grateful to Angela Rinaldi, psychologist, who reviewed the literature on healthy siblings' reactions in chronic illnesses.

List of abbreviations

BI: Barthel Index
BMD: Becker Muscular Dystrophy
DMD: Duchenne Muscular Dystrophy
FPQ: Family Problems Questionnaire
MD: Muscular Dystrophy
MD-CS: Muscular Dystrophy Care Schedule
MDs: Muscular Dystrophies
SNQ: Social Network Questionnaire

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