

Family context in muscular dystrophies: psychosocial aspects and social integration

LORENZA MAGLIANO¹ AND LUISA POLITANO²

¹Department of Psychology, Campania University "Luigi Vanvitelli", Caserta, Italy; ²Cardiomyology and Medical Genetics, Department of Experimental Medicine, Campania University "Luigi Vanvitelli", Naples, Italy

Muscular dystrophies (MDs) are degenerative diseases which may lead to marked functional impairment and reduced life expectancy. Being caregivers of a loved one with MD may be both a rewarding and a demanding experience that may have relevant impact on the quality of life of the whole family. In this short review we summarize the main findings of the first survey on family context in MD in Italy.

The study was carried out on 502 key-relatives of patients suffering from Duchenne, Becker, or Limb-Girdle MD, aged between 4 and 25 years, and attending one of 8 participating Centers, all over 2012. The results revealed that practical difficulties were mainly related to relatives' involvement in helping the patient in moving and in relative's constraints of leisure activities. Furthermore, feelings of loss and perception of patient's condition as having negative effects on the family life were the psychological consequences more frequently complained.

However, despite the difficulties, 88% of the key-relatives acknowledged the caregiving as a positive experience. In fact 94% of the respondents stated they could rely on friends in case of own physical illness, and 88% in case of psychological stress.

Burden was found higher among relatives of patients with lower functional autonomy and longer duration of illness, and among relatives with lower professional and social support. Conversely, the positive aspects of the caregiving were more frequently acknowledged by those who received higher level of professional help and psychological social support.

These results reveal that the caregiving experience has a positive impact on key-relatives quality of life despite the practical demands, and that the support of professionals is essential to help families in identifying the benefits of this experience without denying its difficulties.

Key words: muscular dystrophies, family burden, social network

Introduction

Muscular dystrophies (MDs) are degenerative, rare diseases that lead to muscle strength loss and progressive restriction of functional abilities (1, 2). Although only symptomatic therapies are available for these diseases, improved standards of care have led to a considerable increase in life expectancy (2, 3). Most patients with MD live at home and receive daily assistance from their relatives (4-10).

Difficulties experienced by relatives of patients as a consequence of their caregiving role are commonly referred to as "family burden" and are divided into "practical" and "psychological" burdens (11, 12). Practical burden refers to problems such as disruption of family relationships, constraints in social, leisure, and work activities, and financial difficulties (11-13). Psychological burden describes the reactions that family members experience, e.g. feeling of loss, sadness, tension, and feeling unable to cope with the situation (14-16).

Family involvement may facilitate patients' adaptation to the illness and their clinical response to therapies, but it can lead to high family burden. Practical and psychological consequences of family caregiving have been rarely examined in neuromuscular diseases.

Available data suggest that different pathologies, due to their clinical characteristics and social reactions to them (11), may dictate specific needs for care and require different therapeutic strategies (12, 17-19).

Family involvement in the care of long-term diseases is particularly relevant in Italy, where the national health policy is strongly community oriented (11). In Italy, no study has systematically explored the burden, and social and professional support in key-relatives and healthy sibling of children with MD.

In 2012, a survey on the families of young patients with several forms of MD, including DMD, Becker MD (BMD), and Limb-Girdle MDs (LGMDs) was carried out on a national scale. The project aimed to explore the following aspects: 1) practical and psychological burden in key-relatives; 2) practical and social network support; 3) pattern of care received by the patients and professional support to the caregivers; 4) differences related to type of MD, pattern of care, and geographical areas.

In this invited review, we summarize the main findings from the above mentioned study focusing on psychological benefits, main practical difficulties and social and professional resources (20-22).

Patients and methods

Patients

A total of 502 key relatives of 4-25 year old patients with MDs who were enrolled in 8 specialized Italian centers for MDs participated in the survey (Fig. 1). Patients' selection criteria: age between 4 and 18 years; in charge for at least 6 months; living with at least one relative; not suffering from diseases other than those MD-related. Key-relatives' selection criteria: age between 18 and 80 years; not suffering for illness requiring long-term intensive care; not living with persons suffering from chronic illness but the patient. The study protocol was approved by the Ethics Committee of the Second University of Naples and by the Local Ethics Committee of each participating Center.

Methods

To assess the patient's functional autonomy an *ad hoc* semi-structured interview was developed and used to obtain the Barthel-10 functioning index (23) by the

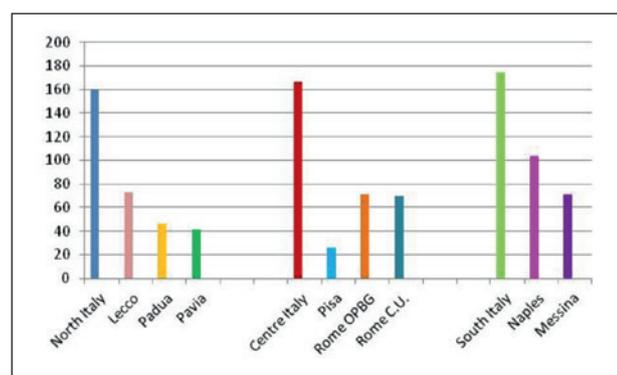


Figure 1. Distribution of the enrolled cases according to the geographical area (North, Center and South Italy). Legend: OPBG: Bambin Gesù Paediatric Hospital; C.U.: Catholic University

key-relative. Family Problems (FPQ) and Social Network (SNQ) Questionnaires were administered to key-relatives in order to focus on the difficulties and resources experienced by the families (11).

Results

Patients

The majority of the patients were male (96%), young (mean age 12.8 (5.6sd), and in school (86%); 66% of them suffered from DMD, 26% from BMD, and 15% from LGMD. Sixty-one percent of patients were ambulant and 39% wheelchair-bound, with a mean level of independence in daily activities, measured by the Barthel Index, of 68.3 (31.3 sd). Most of patients were in drug treatments (73%) and attended rehabilitation programs (67%). Only 72 patients (14%) had received psycho-educational interventions as psychological support (53%) and information on muscular dystrophies' treatments (39%). Moreover, 66% of patients received social/welfare support, mainly economic benefits and 16% school support.

Key-relatives

Most of the key-relatives were mothers (84%) and lived with a partner (88%). Almost half of them (56%) had received higher education and 53% were employed. In the two months preceding the evaluation, key-relatives spent on average 5.7 (4.6sd) daily hours in patient's caregiving. In the previous six months, 31 relatives received psycho-educational interventions including education on clinical and rehabilitative procedures (68%), information on treatments (54%), and psychological support (22%). Of the 55 of relatives (11%) receiving social/welfare support, 46 (84%) were sustained by Family/Patients Associations. As far as the practical consequences of caregiving, the most frequently mentioned difficulties were the neglect of hobbies and free time activities (59%), night awakenings to take care of their patient (45%), and difficulties in work and household activities (45%). Moreover, 35% of relatives stated that they had economic difficulties and 64% reported to have sustained costs for patient's care (doctors/nurses and drugs). Regarding psychological difficulties, 77% of the relatives reported feelings of loss, 74% sadness and/or depression, and 72% worries for the future of other family members.

However, despite the difficulties, 88% of key-relatives acknowledged the caregiving experience as having a positive impact on their lives. In particular, 72% reported changes in life's values, and 18% an increased sense of strength and courage against adversities. Moreover, 94% of the relatives stated they could rely on friends in case

of own physical illness, and 88% in case of psychological stress. Furthermore, 92% felt their friends would help them in case of patient's emergencies, and 97% sure to receive professional help in a crisis situation.

The study revealed that burden was higher among relatives who: a) were unemployed and single; b) had patients not attending school and with DMD; c) had less support by their social network and the professionals. Conversely, the positive aspects of caregiving were more acknowledged by key-relatives who had a higher level of professional help and psychological social support.

Exploring the differences among the geographical areas, the study outlined that the welfare support was more frequently available in Northern Italy, the psycho-educational interventions in Central Italy and the clinical care of cardiologist aspects in Southern Italy.

Discussion

These findings confirm that home management of patients with MDs may be demanding for patients' relatives, especially when social and professional resources are poor and patients' functional abilities decrease (24). On the other hand, these findings highlight how to rely on the various types of support (social network, professionals and welfare) make differences in terms of family resilience and coping strategies (25-30). In particular, the results of this study will be useful for clinicians to better understand the complexity of the caregiving process in muscular dystrophies and for the healthy policy managers to plan an appropriate allocation of resources.

Acknowledgements

The study was supported by a Telethon UILDM grant (Grant n. GUP10002).

We are grateful to the Principal Investigators from the Participating Centers: U. Balottin, IRCCS Fondazione Istituto Neurologico C. Mondino, University of Pavia, Pavia; G.Vita, University of Messina; M. Pane, Università Cattolica del Sacro Cuore, Rome; A.D'Amico, Bambin Gesù, Paediatric Hospital, Rome; C. Angelini, University of Padua, Padua; R. Battini – Fondazione IRCCS Stella Maris, Pisa; M.G. D'Angelo, I.S.E. Medea, IRCCS La Nostra Famiglia, Bosisio Parini (LC), and their coworkers (GUP10002 working group).

References

- Emery AEH. The muscular dystrophies. *Lancet* 2002;359:687-95.
- Bushby K, Finkel R, Birnkrant DJ, et al. Diagnosis and management of Duchenne muscular dystrophy, part 1: diagnosis, and pharmacological and psychosocial management. *Lancet Neurol* 2010;9:77-93.
- Passamano L, Taglia A, Palladino A, et al. Improvement of survival in Duchenne Muscular Dystrophy: retrospective analysis of 835 patients. *Acta Myol* 2012;31:121-5.
- Anderson M, Elliott EJ, Zurynski Y. Australian families living with rare disease: experiences of diagnosis, health services use and needs for psychosocial support. *OJRD* 2013;28:22.
- Baiardini I, Minetti C, Bonifacino S, et al. Quality of life in Duchenne muscular dystrophy: the subjective impact on children and parents. *J Child Neurol* 2011;26:707-13.
- Green JM, Murton FE. Diagnosis of Duchenne muscular dystrophy: parents' experiences and satisfaction. *Child Care Health Dev* 1996;22:113-28.
- Im SH, Lee SC, Moon JH, et al. Quality of life for primary caregivers of muscular dystrophy. Subject description. *Chin Med J (Engl)* 2010;123:452-7.
- Kenneson A, Bobo JK. The effect of caregiving on women in families with Duchenne/Becker muscular dystrophy. *Health Soc Care Community* 2010;18:520-8.
- Ouyang L, Grosse SD, Fox MH, et al. A national profile of health care and family impacts of children with muscular dystrophy and special health care needs in the United States. *J Child Neurol* 2012;27:569-76.
- Pangalila RF, Van den Bos GM, Stam HJ, et al. Subjective caregiver burden of parents of adults with Duchenne muscular dystrophy. *Disabil Rehabil* 2012;34:988-96.
- Magliano L, Fiorillo A, De Rosa C, et al. Family burden in long-term diseases: a comparative study in schizophrenia vs. physical disorders. *Soc Sci Med* 2005;61:313-22.
- Rossi Ferrario S, Zotti AM, Ippoliti M, et al. Caregiving-related needs analysis: a proposed model reflecting current research and socio-political developments. *Heal Soc Care Community* 2003;11:103-10.
- Chen J, Clark M. Family function in families of children with Duchenne muscular dystrophy. *Family and Community Health* 2007;30:296-304.
- Abi Daoud MS, Dooley JM, Gordon KE. Depression in parents of children with Duchenne muscular dystrophy. *Pediatr Neurol* 2004;31:16-9.
- Bothwell JE, Dooley JM, Gordon KE, et al. Duchenne muscular dystrophy-parental perceptions. *Clin Pediatr (Phila)* 2002;41:105-9.
- Larkindale J, Yang W, Hogan PF, et al. Cost of illness for neuromuscular diseases in the United States. *Muscle Nerve* 2014;49:431-8.
- Eccleston C, Palermo TM, Fisher E, et al. Psychological interventions for parents of children and adolescents with chronic illness. In: *Cochrane Database Systematic Review*. Published online: August 15, 2012. DOI: 10.1002/14651858.CD009660.
- Farmer JE, Marien WE, Clark MJ, et al. Primary care supports for children with chronic health conditions: identifying and predicting unmet family needs. *J Pediatr Psychol* 2004;29:355-67.
- Vanasse M, Paré H, Zeller R. Medical and psychosocial considerations in rehabilitation care of childhood neuromuscular diseases. *Handb Clin Neurol* 2013;113:1491-5.
- Magliano L, Patalano M, Sagliocchi A, et al. "I have got something positive out of this situation": psychological benefits of caregiving in relatives of young people with muscular dystrophy. *J Neurol* 2014;261:188-95.
- Magliano L, Patalano M, Sagliocchi A, et al. The condition of the families of children and young adults with muscular dystrophies in Italy. *Muscle Nerve* 2015;52:13-21.
- Magliano L, D'Angelo MG, Vita G, et al. Psychological and prac-

- tical difficulties among parents and healthy siblings of children with Duchenne vs. Becker muscular dystrophy: an Italian comparative study. *Acta Myol* 2014;33:136-43.
23. Mahoney F, Barthel D. Functional evaluation: the Barthel Index. *Md State Med J* 1965;14:61-5.
 24. Magliano L, Fiorillo A, Malangone C, et al. Social network in long-term diseases: a comparative study in relatives of persons with schizophrenia and physical illnesses versus a sample from the general population. *Soc Sci Med* 2006;62:1392-402.
 25. Cohen C, Colantonio A, Vernich L. Positive aspects of caregiving: rounding out the caregiver experience. *Int J Geriatr Psychiatry* 2002;17:184-8.
 26. Green SE. "We're tired, not sad": benefits and burdens of mothering a child with a disability. *Soc Sci Med* 2007;64:150-63.
 27. Samson A, Tomiak E, Dimillo J, et al. The lived experience of hope among parents of a child with Duchenne muscular dystrophy: perceiving the human being beyond the illness. *Chronic Illn* 2009;5:103-14.
 28. Hodges L, Dibb B. Social comparison within self-help groups: views of parents of children with Duchenne muscular dystrophy. *J Health Psychology* 2010;15:483-92.
 29. Greeff AP, Vansteenwegen A, Gillard J. Current issues resilience in families living with a child with a physical disability. *Rehabilitation Nursing* 2012;37:97-104.
 30. Savundranayagam MY. Receiving while giving: the differential roles of receiving help and satisfaction with help on caregiver rewards among spouses and adult-children. *Int J Geriatr Psychiatry* 2013;doi: 10.1002/gps.3967.