

**A 10-YEAR LITERATURE REVIEW OF FAMILY CAREGIVING FOR MOTOR
NEURONE DISEASE: MOVING FROM CAREGIVER BURDEN STUDIES TO
PALLIATIVE CARE INTERVENTIONS**

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Abstract:

Background: There is growing awareness that different terminal diseases translate into different family caregiver experiences, and the palliative and supportive care needs of these families are both similar and unique. Family members caring for people with motor neurone disease (MND) may experience exceptional strain due to the usually rapid and progressive nature of this terminal illness.

Aim: The purpose of this review is to synthesize contemporary research and provide a comprehensive summary of findings relevant to MND family caregivers, as well as highlight some of the suggested interventions to alleviate burden and improve quality of life for this group.

Design: We conducted a comprehensive review of empirical research on family caregiving for people with MND in peer-reviewed journals published in English, January 2000 - April 2011. Fifty-nine studies met the inclusion criteria.

Results: This comprehensive literature review was consistent with previous research documenting the substantial burden and distress experienced by MND family caregivers, and revealed important points in the trajectory of care that have the potential for negative effects. The diagnosis experience, assisted ventilation, cognitive changes, and end of life decision making, create challenges within a short time. This review has also implicated the need for improvements in access to palliative care services and highlighted the absence of interventions to improve care.

Conclusions: Caregiver burden and quality of life studies on MND family caregivers have so far dominated the research landscape. The focus needs to be on developing interventions that provide direct practical and psychosocial supports for MND caregivers.

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INTRODUCTION

Motor neurone disease (MND) refers to four disorders involving the degeneration of motor neurones controlling voluntary muscles that enable people to move, speak, breathe and swallow. Amyotrophic lateral sclerosis (ALS) is the most common type and the two terms MND and ALS are used synonymously and vary by country. The person with MND endures progressive paralysis, and death is most often caused by respiratory failure (1). The cause is unknown and there is no known cure (2). The median life expectancy is two to four years after diagnosis (3). While presenting symptoms and disease course vary, MND is always progressive with no periods of remission (4). Symptoms include muscle weakness and atrophy, difficulty breathing and swallowing, speech problems, emotional lability, and pain (5). The supportive and palliative care goals for people with MND include maintaining physical function and independence for as long as possible, managing pain and other physical symptoms, facilitating communication about end of life issues, and providing psychosocial support for the person and their family throughout the disease (5-7). Though MND is relatively uncommon, with a prevalence of 7 in 100,000 (8) the associated direct (e.g. health care, expensive equipment) and indirect (lost productivity and income) costs and intangible losses (independence, quality of life) all contribute to the high cost of this illness to society (9).

In general, end-of-life caregiving by families is demanding and associated with morbidity and mortality (10-12). Family members caring for those with MND may experience exceptional strain due to the usually rapid and progressive nature of this terminal

illness, coupled with the particularly debilitating effects of MND (13). Awareness has been mounting about the specific difficulties relating to caring for a person with MND and the palliative and supportive care needs of these families (14, 15). An understanding of the disease trajectory of MND (16), the common challenges faced by MND family caregivers, and how the available support to these family caregivers translates into different caregiver experiences (17) are all highlighting how providing family care for a person with MND is both similar to and unique from family caregiving in other terminal disease populations.

PURPOSE

The purpose of this review is to synthesize contemporary research and provide a comprehensive and up to date summary of findings relevant to family caregivers of people with MND concerning their experiences, burden and quality of life issues, and service needs. Moreover, this review highlights the interventions suggested by the research to alleviate burden and improve the quality of life of MND family caregivers.

Published research from 2000 through April 2011 on family caregiving for people with MND is reviewed. Only one previous review has been published in this area by Mockford, et al, for the period 1994-2004, encompassing 30 studies (18). The main findings of this review indicated that the mental and physical health of caregivers were at risk without improved access to resources and support. Two themes emerged from the review: 1) the impacts on MND carers and 2) service needs and provision.

Since 2004, an additional 43 studies have been published in peer-reviewed journals on a broad range of issues confronting family caregivers of people with MND. We update Mockford's review with the most current research on MND family caregiver impacts and service needs. Sixteen studies are common to both reviews. In addition, our review adds a third dimension: the unique challenges faced during MND family caregiving as a result of the

disease trajectory. These challenges encompass the impact of the diagnosis experience on MND family caregivers, the ongoing difficulties in obtaining adequate information and access to services from health care providers, the toll on family carers when cognitive and neurobehavioral changes associated with MND are encountered, the considerable burden associated with the decision and transition to assisted ventilation, caregiving in the last months of life, and how all of these factors impact on bereavement.

METHODS

We conducted a comprehensive review of empirical research on family caregiving for people with MND published between January 2000 and April 2011. This 10-year period was examined in order to assess the most recent contributions to this field of research and to examine whether there were identifiable and distinct aspects of MND family caregiving.

Inclusion/exclusion criteria

The population addressed is family caregivers who provide care for someone diagnosed with MND, although a definition of “family caregiving” was not applied in the articles. Therefore, we included all articles in which family members or partners were involved in the direct or indirect care of someone with MND. We excluded studies that focused on family carers’ proxy accounts of patient care or symptoms, as well as those documenting instrument development and validation. Only empirical, peer-reviewed journal articles published in English were included. Qualitative, quantitative and mixed methods studies were considered. Exclusions were not made on the basis of assessments of the rigour, validity, and reliability of studies and their findings, as would be typical in a systematic review. Given that most studies on caregiving at the end of life fell into evidence levels III (non-randomised comparative studies) and the limitations of the study designs, it was

difficult to grade using traditional taxonomies for levels of evidence for systematic reviews (19).

Search Strategy

The databases searched were Global Health, Medline, ProQuest Health and Medical Complete, PsychINFO, Science Direct, CINAHL (EBSCO), and Current Contents Connect. We used any combination of the following search terms: palliative care, supportive care, end of life care AND carer(s), caregiver(s), family caregiver(s), family carer(s), AND motor neuron(e) disease, amyotrophic lateral sclerosis, neurodegenerative disease. To augment the computer-based search, a manual search of the reference lists of all retrieved articles was conducted.

RESULTS

The electronic search identified more 1625 articles in all databases. After excluding duplicates and studies where caregiving for those with MND was not a central or partial focus, 216 abstracts were identified for review. Of these, a total of 59 studies met the inclusion criteria (Figure 1). A summary of the main features of the studies reviewed, including locations, sample size, methodology, and primary focus are presented in Table 1. More details on the key characteristics of the reviewed studies are presented in Table 2.

Conceptual and Definitional Issues

Many studies did not clearly define key concepts, such as “caregiver.” Thirty-three (56%) articles did not define it at all, and others used a variety of definitions such as “the primary caregiver or the person most or primarily involved in patient care” (12%). The terms “end of life care” and “palliative /supportive care” were not defined or poorly defined in all the studies, making it difficult to determine the nature of the care being discussed. While

most studies addressed “caregiver burden” and/or “quality of life,” few defined these terms. In the majority of the studies, the care setting where caregiving was being provided was also not stated or clearly defined making it difficult to distinguish the setting of care in many articles. Close inspection of the recruitment procedure and findings suggested that people with MND received primarily home-based care.

STUDIES REVIEWED

Key findings reported in the literature are grouped into three major themes: 1) the unique experiences of family caregiving for someone with MND, with a focus on caregiver time, cognitive and neurobehavioral decline, assisted ventilation, caregiving in the last months of life, requests for hastened death and bereavement; 2) caregiver burden and quality of life issues; and 3) identified service needs.

The unique experiences of caring for someone with MND

A family caregiver for a person with MND quickly encounters life changes of great magnitude. MND typically involves a rapid functional decline with a short disease trajectory; 50% die within 3 years of symptom onset (20). During this time, people with MND lose the ability to move, eat, breathe, and speak, necessitating transitions to wheelchairs and the use of other mobility aids, Percutaneous Enteral Gastronomy (PEG) feeding, the use of assistive communication devices, and the use of assisted breathing devices(Non-Invasive Positive-Pressure Ventilation [NIPPV] and/or tracheostomy and Long Term Mechanical Ventilation [LTMV]) (21). Thus, caregivers face daily changes, as well as long term adjustments (22). Stresses and challenges include: dealing with swallowing and eating difficulties (23); worries about the illness progression and the well-being of family (23, 24); changes in family roles (22, 25); loss of a sexual relationship between spouse caregivers and people with MND (21,

26); loss of a reciprocal relationship with spouse caregivers (27); and being blamed by other family members for inadequate care (28).

Caregiver time

Over time, people with MND require increasing levels of assistance with nearly all aspects of daily living (13). In an Italian study, it was reported that caregiver time for people with MND is highly related to overall disability and ranged from 5 hours per day for those with mild disability to 15 hours per day for those with severe impairment. Losing the ability to walk had the greatest impact on caregiving time, while the need for enteral nutrition or assisted ventilation had marginal impact. A progressive increase was observed in time spent on toileting, bathing, dressing, administering medications, and feeding, corresponding with level of disability (29).

Cognitive and neurobehavioral decline

Up to fifty percent of people with MND will develop neurobehavioral symptoms, from a subtle cognitive decline to serious impairment (30). Apathy, executive dysfunction and disinhibition are the most common neurobehavioral symptoms of patients reported by caregivers (30, 31). In about 15% of cases, people with MND will develop frontotemporal dementia which presents additional challenging neurobehavioral symptoms for family caregivers including irritability, impulsivity, loss of insight, self-centeredness and lack of empathy, aggression, food cravings, and obsessions and/or compulsive behaviors (30). A German study found that burden is markedly higher in MND caregivers who report encountering “problem-behavior” (32). Cognitive changes have a profound impact on caregivers, sharply increasing depression and burden and negatively impacting their quality of life (31).

Assisted ventilation

Respiratory muscle weakness increases over time and people with MND begin to experience hypoventilation (33). Five studies looked at how the symptom of breathlessness and subsequent assisted ventilation, if chosen, affected caregiver burden and quality of life. A study in the U.K. found caregivers generally feel helpless when confronted with breathlessness and ill-prepared to cope, causing anxiety, depression, and distress (34). Both NIPPV and LTMV increase survival time for people with MND, leading to an extended period of caregiving (33, 35). Studies in the U.S., Japan and Germany point to the particular role of LTMV in caregiver burden, stress and distress (21, 35, 36). Burden was reported in relation to how LTMV caregivers often become acutely aware of their responsibility for the survival of their family member, from managing the tracheostomy and suctioning the airway, to managing equipment and arranging back-up power sources. Because of these responsibilities, family caregivers of people on LTMV often give up all but essential outside activities and relinquish their personal lives (35, 36).

End-of-life caregiving and requests for hastened death

At the end of life, as family caregivers lose the ability to interact with their loved ones, they begin to struggle with meaning and feel powerless (36). Pagnini (37) found that, as people with MND lost function, their caregivers exhibited increased symptoms of somatic depression such as reduced quality of sleep and appetite. There is some data to indicate that people with MND and their family caregivers do not always hold the same attitudes or beliefs about care (38), or agree about treatment course or end-of-life decision making (39). People with MND may refuse life-sustaining treatments, such as tracheostomy or assisted ventilation (21), or may have interest in hastening death, all of which can cause distress for their caregivers (40).

Bereavement

MND continues to affect families after death, despite the end to caregiving. In bereavement, MND caregivers experienced lasting emotional impacts, including sadness, fear, frustration, hatred and anger, depression, and, for some, hopelessness (41). An Australian study demonstrated that MND family caregivers are at risk of prolonged grief (42). While there is a strong recognition for the value of grief and bereavement support, this support is often inadequate (41, 42). In a 2001 Canadian study, 37% of caregivers were coping poorly or very poorly, sometimes years after death (28). Martin and Turnbull (28) also found lasting financial hardship reported by 22% to 26% of bereaved families.

Caregiver Burden and Quality of Life

MND caregiver burden and quality of life (QOL) were explored within 40 articles in this review, all of which documented the considerable burden attached to care provision (43, 44). Burden was most frequently reported in the domain of time-dependence, reflecting the lack of time for oneself as the main reason for caregiver strain (13, 15, 37, 45). Generally negative outcomes on MND caregiver QOL were documented (44, 46-48). Caregiver QOL appears unrelated to the severity of MND, patient QOL, or the level of care (40, 49, 50). In a longitudinal study, Gauthier (15) found caregiver emotional burden and total burden increased over time, but QOL remained stable.

Mitigating factors associated with caregiver burden and quality of life

Caregiver burden increases as patient function declines (15, 37, 51), but caregiver QOL appears dependent on factors other than the physical decline of the care recipient (49) such as religiousness (52). Caregiver burden may be mitigated through positive social support (26, 37, 49, 53, 54), Studies in the U.K. and Australia identified how high

satisfaction with social support helped mitigate potentially negative effects of MND on quality of life and the marital relationship between spouses and caregivers (26, 53). Several studies (22, 38, 55, 56) indicated that family caregivers need a confidant to mitigate isolation. Yet MND caregivers, particularly those providing prolonged care, face challenges seeking and maintaining social support, interaction, and help from informal networks (13, 54, 57). MND caregivers tend to have reduced social contacts, social network size and quality due to caregiving (39, 54, 58), which may contribute to social isolation and perceived burden (54). There are indications that female MND caregivers may experience greater burden than men (13, 31, 32, 53, 59).

Caregiver physical concerns

MND caregivers commonly expressed concerns about their own health (37). The most reported physical health concerns among MND caregivers include physical exhaustion or fatigue (15, 27, 40, 57, 60), followed by insomnia and interrupted sleep (46, 57, 61, 62). MND caregivers may have self-care difficulties due to a lack of time for their own needs (60).

Depression and Anxiety

The most commonly documented mental and psychological health symptoms and issues among MND caregivers included depression (13, 15, 39-41, 45, 46, 60, 63, 64); psychological distress (15, 40, 50, 57, 60); and anxiety (27, 41, 46, 57, 62, 63, 65). Depression rates may be influenced by culture or ethnicity. One study using a Japanese sample of MND caregivers reported extremely high rates of depression (61%) (45); in contrast, two studies of American MND caregivers report low rates (19% and 23%) even when patients were severely disabled (40, 60).

Depression tends to increase over time (15, 51) in connection with multiple losses and changes faced by MND family caregivers (27), as well as caregiving intensity and supervision time (45); perceptions of the patient as unsupportive or critical (60); household income (45); burden (15); and the use of avoidance coping strategies rather than problem-solving (60). Caregiver depression may be related to patient depression (13, 40). Rabkin et al. (60) indicated that psychological distress, rather than psychopathology (e.g. clinical depression), was more reflective of the circumstances of caregivers of late-stage MND patients.

Only one study conducted in Italy formally assessed anxiety in MND caregivers, reporting prevalence rates from 71 to 79% (65). MND caregiver anxiety may be associated with the levels of the patient's disability (38, 44); or may arise from being unable to make plans on a daily basis, due to the uncertainty of disease trajectory (27).

Service needs among caregivers of people with MND

Information and communication

Communication difficulties with health professionals over obtaining adequate information about diagnosis, health status, prognosis, trajectory, and treatment options were identified in several studies (22, 25, 38, 55, 62, 66-68). Caregivers in Australia and the U.K. were distressed when it took a long time to obtain a diagnosis, and when medical specialists lacked empathy and communication skills (68, 69). Diagnostic experiences can be particularly stressful for caregivers (70). However, studies suggested that communication at this time could be improved (38, 42, 55, 71). McClusky et al., (71) found the average time spent imparting the diagnosis in a U.S. study was 30 minutes, and while 65% of doctors identified MND as a terminal disease, only 11% discussed palliative care options. Four qualitative studies describe the poor experiences of patients and family caregivers at the time

of diagnosis, including receiving the news over the phone in a short, business-like phone call with minimal information (19, 55). In cases where diagnosis is relayed poorly, there can be considerable emotional consequences for family caregivers that may linger into bereavement (42).

Even when caregivers were largely satisfied with how the diagnosis was communicated, they also expressed barriers to having their information needs met, and presented a need from the beginning for more general information about MND prognosis, as well as information about current MND research, therapies that slow down MND progression, potential cures, MND outcomes, and information on ventilation (22, 36, 62, 66). Access to and information about resources and equipment was another identified need (25, 38, 46, 62), as is help with family communication (46, 47).

Identified needs and service gaps in palliative/supportive care

Assistance for caregivers with several aspects of patient care needs and symptom management were indicated in several Australian studies, as well as related needs for home support, home care and respite, and financial assistance (28, 46, 47, 69). Access to experts from a variety of agencies (62) and access to community rehabilitation are identified needs (46, 47), but respite care was singled out as the most important service that would enable caregivers to continue providing care (72).

Other needs include direct help or guidance with practical/medical aspects of care (i.e., symptom and medication management, technological tasks) (25); personal care needs (dressing, washing and showering) (25, 46, 72); household and instrumental tasks (25, 72). Persistent patient breathlessness at the end of life has been identified as one of the most difficult symptoms for caregivers (including MND caregivers) to deal with (34).

As MND has an identifiable terminal stage from the start of diagnosis, a palliative care approach for MND has been emphasized (7, 46, 47). Palliative care approaches, including symptom management, supportive services, counselling and family care plans, are relevant for MND. They can also help prepare MND caregivers for the physical and emotional decline in people with MND (62). Nonetheless, there is evidence that when palliative care is available, both people with MND and their caregivers find it difficult to access these services (20, 73). In a Western Australian study, it was found that less than 10% of people who died of non-malignant diseases had accessed specialist palliative care services, compared with 66% of people who died of cancer (74). Thus, there appears to be a need for increased understanding and application of a palliative care approach to care delivery for those with MND and their caregivers.

DISCUSSION AND CONCLUSIONS

This review summarized the literature between 2000 and early 2011. While caregiver burden and quality of life studies have dominated the research and documented the substantial distress experienced by MND family caregivers, more studies are now emerging that provide guidance about how to provide optimal palliative and supportive care to people with MND and their family caregivers. Important milestones in care have been identified, such as diagnosis, accessing information and services, cognitive and behavioral changes that can occur in MND, the decision to receive assisted ventilation, end of life decision-making, and bereavement. However, the development and testing of interventions focused on providing direct supports and outcomes for MND caregivers appears to be lacking.

Limitations

Many of the study methodologies have limitations that weaken the validity of the findings, including: small sample sizes and non-random samples. Most studies of

psychological distress in MND caregivers were further limited by non-standardised instruments and caregiver self-reports (rather than systematic clinical approaches). Female caregivers are overrepresented in the studies. Caregivers at the end of life were underrepresented as people with MND who are more severely impaired are harder to recruit and their caregivers are less likely to participate because they are coping with increased disease burden. Also, due to ethical reasons, some studies excluded caregivers who were caring for patients at the last stage of MND (e.g. 55). This limitation may also arise from gate-keeping, where health care practitioners may seek to exclude patients they see as vulnerable from participating in research (46, 47).

Limitations of this review include the focus on studies published in English; relevant studies in other languages may exist. A lack of generalisation to other ethnic/cultural groups and to those from non-English-speaking backgrounds is also indicated. Additionally, the studies were not assessed for the quality of the research design and we did not fully consider the psychometric properties of various rating scales and instruments used by the researchers. Except for one large-scale national study (46, 47) and one European study (44), most studies were local, single-site research, involving mostly non-representative samples with a potential for bias. Given the diverse terminologies, differing objectives and a range of measures used in MND caregiving studies, there are considerable challenges in attempts to analyse and compare important constructs (such as caregiver burden, psychological distress, QOL, etc.) among the studies. Nevertheless, there is enough consistency in the findings to explore the implications for service provision and inform future research.

Implications for service provision

While studies have continued to document evidence of burden and distress in MND family caregivers, this evidence is rarely translated into improvements in practice through the

development of interventions. Six especially challenging points in the caregiving trajectory emerged from the recent literature: the diagnosis, obtaining information and access to health care services, the decision and transition to assisted ventilation, encountering neurobehavioral changes in the patient, caregiving and decision making in the last months of life, and bereavement. The types of interventions suggested in the literature reviewed to support MND family caregivers can be grouped into three broad and overlapping areas: 1) providing direct support to family caregivers of people with MND to meet their practical and emotional needs; 2) providing education and training to health care providers in order that they may better meet the emotional and practical needs of people with MND and their family caregivers; and 3) providing interventions which support the existential and spiritual concerns of both people with MND and family caregivers. To date, there are no intervention studies that assessed direct supports and outcomes for MND caregivers. However, certain recommendations for future interventions could be gleaned from many of the studies.

Providing supportive services and addressing caregivers' support networks

As there is a clear correlation between quality of life and the amount of support experienced by MND caregivers (37), interventions that bolster support were the most commonly implicated by the studies reviewed. This included interventions in the caregiving situation through: providing paid assistance for home help services(72); helping caregivers both socially connect with and mobilize their informal networks (32, 58); connect with community support groups (13); providing direct psychological support (60); and targeted delivery of psycho-educational programs (50, 63, 64).

The quality of life of both people with MND and their family caregivers appears primarily dependent on relationship and psychosocial issues, and the wellbeing of both parties is linked. Counselling interventions can support the patient and caregiver relationship

(24). At least two studies indicated that spouse caregivers of people with MND are concerned with the loss of their sexual relationships, and interventions should be developed to address this often overlooked area (21, 26).

Emotional support programs for both the family caregiver and person with MND are indicated to mitigate the diagnosis experience (68). For caregivers who encounter neurobehavioral issues in care recipients, support interventions that offer problem-solving strategies and which provide emotional support are needed (31). As the patient approaches the end-of-life, counselling interventions are needed to address distress, anxiety and somatic expressions of depression (37). The identification of risk factors for prolonged grief is indicated for the MND family caregiver, and more tailored bereavement support programs should be developed (42).

Education and training for health care providers

Several studies indicated that professional health care providers need more training to improve their communication of the diagnosis to the person with MND and their family, to provide useful information to families after diagnosis (1, 66), and to effectively communicate about end of life care preferences (75). Professional education and training programs can enhance the understanding of MND by service providers (20), as well as their understanding of a palliative approach for people with MND, ability to promote dignity and quality of life and better communication with MND patients and their caregivers (76).

Interventions supporting existential and spiritual concerns

The development of spiritually-based interventions have been indicated by several studies to potentially improve MND caregiver's quality of life (49, 50, 52, 77). An Italian study has indicated that interventions that improve existential wellbeing in people with MND

will also improve the psychological wellbeing of their family caregivers (37). Dignity Therapy (78), which focuses on maintaining hope and a sense of meaning in the patient, may reduce requests for a hastened death at the end of life, indirectly reducing caregiver distress (79). Dignity Therapy has also been shown to moderate the bereavement experience of family members, indicating another possible benefit to MND caregivers (80). These types of interventions and others may be developed and evaluated for future use in MND caregivers.

Conclusion

Research findings have been consistent in documenting the substantial burden and distress experienced by MND family caregivers. Numerous suggestions have emerged through the research for the development of interventions and supportive services to improve the psychological wellbeing and quality of life for these caregivers. The research literature has also documented the need for improved access to palliative care services and made suggestions for interventions to improve care for both people with MND and their family caregivers. Meanwhile, there appears to be a lack of research into the development of such interventions which will alleviate the burden on MND family carers and improve care for patients.

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Table 1: Summary of main features of MND family caregiver studies

Characteristic	n=59
Country	
Australia	13
US	13
UK	13
Italy	10
Japan	3
Sweden	2
Canada	2
Germany	2
Multi-national (15 European countries)	1
Primary Focus	
Caregiver burden and quality of life	38
Service use	8
Supportive/palliative care	7
Caregiver needs	6
Studies which also included a secondary focus	18
Methodology	
Quantitative	39
Qualitative	17
Mixed Method	3
Disease studied	
Only MND	47
MND, Huntington Disease, Parkinson Disease, Multiple Sclerosis	7
MND, Parkinson Disease, Multiple System Atrophy, Spinocerebellar Degeneration	2
MND, Muscular Dystrophy	1
MND, Chronic Obstructive Pulmonary Disease, heart failure, lung cancer	1
MND, MS, cancer, stroke, cardiovascular disease, respiratory disease, diabetes, arthritis	1
Family caregiver type	
Spouse/Partners only	9
Family caregivers including spouse caregivers	25
Any unpaid family or friend caregivers	12
Sample includes paid caregivers along with family caregivers	3
Insufficient information or not defined	10
Sample size	
Less than 25	21
25-49	12
50-99	17
100-199	5
200-299	0
300-399	2
400 and more	2

