Motor Neurone Disease and Palliative Care

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Abstract
This paper explores the progression of motor neurone disease (MND) in order to outline the complex needs and care requirements with respect to symptoms, suffering, grief and loss, and the potential for early referral to palliative care to ameliorate the impact of the disease. MND is a neurological disorder affecting motor neurones and the muscles under voluntary control, leading to deteriorating function of mobility, swallowing, speech and respiration. The person with MND has to constantly adapt to multiple losses and increasing paralysis, which affects them emotionally, physically, psychologically and spiritually. Ultimately they will die from respiratory failure. Palliative care can provide symptom management, comfort and care to the person, their carers and family, and help to ameliorate the distress and impact of a devastating illness. Their multidisciplinary team is able to provide holistic care and resources that can enhance the quality of life and assist with grief and loss as well as provide bereavement support for those affected.

Development of the role of MND Shared Care Worker
In 2007 the Department of Human Services, in conjunction with Motor Neurone Disease Victoria (MNDV), commissioned a project to assist people with motor neurone disease (MND) to receive access to palliative care services and to assist palliative care services to manage and support people living with MND. The report found that palliative care workers did not feel confident in their level of knowledge with respect to MND and that people with MND did not fully understand the services available through palliative care. When people with MND were in in-patient units, due to their high and complex care needs, staff were affected by reduced staff time and resources for patient care. Following this report, shared care worker positions were implemented across Victoria to be located in palliative care agencies in order to promote early referral, support, development and delivery of education to service providers and coordination between service providers and families. There was an establishment of guidelines and a suitable mechanism to provide supplementary funding for in-patient and community palliative care services to meet the identified high and ongoing care needs of people with MND (MHACSD, 2008).

The Impact of Motor Neurone Disease
Motor neurone disease, amyotrophic lateral sclerosis (ALS) or Lou Gehrig’s disease (as it is sometimes called in the USA), is used to describe a disease where there is damage to motor neurones. Motor neurones are nerve cells that control the movement of muscles that are under voluntary control. These include all the muscles of the trunk and limbs and of speech and swallowing. Without activation, muscles gradually weaken and waste, resulting in paralysis. The initial symptoms of MND may be weakness in the hands or feet, swallowing difficulties or slurred speech. Muscle twitching and/or cramps may also occur. People with MND have their sensory systems intact but half may experience some form of cognitive impact ranging from minor, in most cases, to frontotemporal dementia in approximately 5–10%. (This figure is growing as more research is carried out compared to the Rocha et al. levels of 3.5% in 2005.) The disease is mostly in the over-55 age group but some young people have developed the disease (Rocha et al., 2005; MNDcare, 2011).

In Victoria the prevalence of MND is approximately 250 people, with an annual incidence rate of 100 new cases. It is estimated that for every person diagnosed with the disease, another 14 family members and close friends will be affected. At September 2007, 242 people with MND were registered with MNDV. In the year ended June 2007, MNDV worked with 364 people with MND, with 140 new registrations and 110 deaths reported. People with MND and their families and carers often suffer considerable psychological and emotional distress before the diagnosis due to worrying symptoms, difficulty identifying the cause, and a protracted period of investigation (MHACSD, 2008).

Each person with MND is affected differently in terms of their initial symptoms, the rate and pattern of disease progression and the length of time to death. There are no remissions and there is no cure. Riluzole, the drug therapy for MND, will only slow the progression of the disease by months. The average time between diagnosis and death is two to three years, with very few people surviving more than five years. Progression of MND is rapid, quickly creating high levels of disability and the consequent need for support, including assistance with feeding, communication, movement, transferring, toileting, and other personal care activities. MND has an impact on all activities of daily living (Oliver, 2002).

The progression of MND means that the level of disability changes over months and weeks rather than years. Health care professionals who are unaccustomed to managing MND may not recognise the significance of progressive symptoms. This can lead to unnecessary hardship for those with the condition. The speed of progression creates problems of adjustment for people who have MND, an escalating burden on carers and families, and challenges for service providers and their staff who are charged with meeting the variable and complex care needs (MHACSD, 2008).

It is critically important that health professionals are able to communicate, balancing both hope and empathy and honesty to avoid secondary traumatisation around any diagnostic and care discussions with
difficult topics, such as advanced care plans and the death and dying process (Clayton et al., 2007). Brayford (1992) wrote about the challenges of facing metastatic prostate cancer:

To the typical physician ... my illness is a routine incident in his rounds while for me it’s the crisis of my life. I would feel better if I had a doctor who at least perceived this incongruity ... I just wish he would ... give me his whole mind just once, be bonded with me for a brief space, survey my soul as well as my flesh, to get at my illness, for each man is ill in his own way. (p. 43)

Sickness is one thing but to have your whole identity and personhood subsumed by it creates despair of body, mind and soul (Chochinov, 2007, p. 335).

**Motor Neurone Disease and Palliative Care**

The field of palliative care has been experiencing a shift into caring for those with nonmalignant, life-limiting conditions. Palliative care aims to provide holistic care, manage symptoms and sustain quality of life for the duration of illness through the provision of specialist palliative care services that encompass physical, psychological, emotional and spiritual dimensions. The person with MND benefits greatly from early referral to palliative care where a palliative approach is best taken from diagnosis. For the person with MND, an early referral permits the creation of relationships with health care professionals. This is important as ongoing conversations around advance care planning and constant adjustment to deteriorating health and function need to be revisited on a regular basis. Early referral assists communication when speech is still intact (Clayton et al., 2007, p. 91).

The complex care needs at a physical level include respiratory function, mobility, hydration and nutrition, secretion, bowel and pain management, and communication. Although sensory messages are intact, there may be little or no pain or discomfort. However, pain can occur due to cramps in muscles and legs, spasticity, pain and stiffness in joints and excessive strain on ligaments and soft tissues. The shoulder joint is often affected. Pain in muscles is due to loss of normal postural control and dependent oedema, impaired circulation and skin pressure can also contribute. In addition to physical pain, there is considerable suffering as functionality is reduced and independence removed as the disease progresses insidiously.

Barbato describes the difficulty of understanding the suffering of the dying and their capacity to communicate this type of often unrecognised pain. The richness of metaphor and image to express the experience and emotion may help when words fail, hence the importance of finding avenues for patients and families and carers to express their distress (Barbato, 2006).

The impact of the disease process. Carers may find themselves becoming the manager of household tasks previously carried out by their loved ones. Finances are affected by growing disability and incapacity for work. Social networks are altered – sometimes dramatically – as people find their discomfort with the disability and the slow and complex requirements for communication make visiting uncomfortable. Avoidance of the person with MND may follow. In order for care to take place at home, the carer assumes the role of nurse, companion, problem-solver, and coordinator.

The person with MND and their family are dealing with multiple losses in a progressive and challenging disease process. Carers may find themselves becoming the manager of household tasks previously carried out by their loved ones. Finances are affected by growing disability and incapacity for work. Social networks are altered – sometimes dramatically – as people find their discomfort with the disability and the slow and complex requirements for communication make visiting uncomfortable. Avoidance of the person with MND may follow. In order for care to take place at home, the carer assumes the role of nurse, companion, problem-solver, and coordinator.
of medical appointments and the multiplicity of visits from health professionals, local government and welfare agencies, sometimes while continuing to juggle child care, work and family demands. Carer burden is great and they need high levels of support and appropriate and timely respite. Respite and support can be provided by palliative care. Needless to say there is a great adjustment to the relationship when moving from partner/spouse to full-time carer. Carers need to have support and someone that they can communicate honestly with, and seek to, have some of their own needs (particularly sleep) met in order to continue to be able to care with minimal impact on their own health.

Grief is a process of adjustment to loss and has physical, cognitive, emotional and spiritual dimensions, (Ray, 2006). The person with MND is gradually transformed from a state of independence and agency to one of complete disability. Although many people with MND have communication aids for assistance, they will still experience high levels of distress and frustration when attempting to communicate. It is imperative that health professionals communicate with one another with respect to care requirements in order to avoid exhausting and frustrating repetitive questions. In addition, any emotion is difficult to express in the face of lack of speech and the capacity to express body language. Anger is a common response to such difficulties. Naturally enough, the carer knows exactly what their loved one requires much of the time due to routines and close proximity, so the person with MND becomes highly dependent on their carer and they often share a “system” that serves them both well.

Health care professionals who lack the knowledge and familiarity of MND care may find themselves overwhelmed. Emotionally they may feel helpless against the distress that they feel on witnessing the impact of MND on their patients. They experience their own frustration due to the inability of the person with MND not wishing to take, what health professionals view as, the very necessary interventions and equipment. This is often because the person with MND is struggling to come to terms with their illness and its life-limiting outcome and is not psychologically or emotionally ready to embrace these realities or conversations. Health care professionals inside palliative care and in external agencies need to communicate well and carefully around these issues to avoid negatively affecting their clients.

**Issues for Carers**

The provision of respite to carers is an essential component of care. It can be difficult to help the carer take up the offer of respite and this is sometimes because they feel they should be able to manage without it. They may feel guilt or anxiety about the quality of care their loved one may receive in their absence. The person with MND may be reluctant to go into respite for fear that they will not have the same level of care, support or understanding as they receive from their loved one/carer. Given their high level of dependency this is a real and present fear. When health care professionals are able to demonstrate competent, confident care of the person with MND and provide opportunities to discuss issues that may be affecting choices around respite, there may be a better uptake of respite by carers. Respite is of little value if the carer continues to remain at the bedside while their loved one is in a facility, or if they continue to be woken in the night to attend to elements of care that respite workers are not able to do.

The level of complexity in care needs means that sometimes there is an inadequate level of skilled support and practical help that would enable carers to rest and recuperate (Whitehead et al., 2011, p. 372).

Carers may experience guilt and self-criticism after bereavement about the level of care that they gave. Even though death is anticipated, many carers are fearful of the event and how they will cope (Whitehead et al., 2011, p. 371).

Complicating the clinical picture is the fact that people with MND may have emotional latency when there is upper motor neurone involvement. There may also be evidence of frontotemporal dementia, which has both behavioural, cognitive and language issues. Symptoms can include: disinhibition (inappropriate behaviour and loss of social tact), distractibility (lack of concentration and persistence), and repetitive behaviour (checking and rechecking, fixed and ritualistic behaviour). People suffering from frontotemporal dementia may also exhibit inflexible thinking and impaired judgement, lack of insight and loss of empathy and detachment. This means they may have little awareness of their carers needs or the impact of their behaviour. They may lack interest and motivation for tasks and activities and be somewhat compulsive as well as obsessive with certain foods. There may be accompanying emotional changes, such as depression, anxiety, paranoia, somatic preoccupation and lack of affect. Although there may be difficulties with speech from a mechanical point of view, this can be exacerbated by the frontotemporal dementia and communication and language issues.

**Assessment of Mental Health in MND Patients And Carers**

It is difficult to sort out how the person with MND may be in terms of their mental health. Are they experiencing grief responses in the face of devastating and cumulative losses or are they clinically depressed? Do they have symptoms of frontotemporal dementia or emotional lability? Perhaps they are responding to particular stressors at this point in time. These are important clinical distinctions and should be assessed thoroughly. Discussions with carers about their experience of their loved one are important to distinguish premorbid personality and presentation and current state of mind and behaviour. If frontotemporal dementia is suspected, referral should be made to a neuropsychologist for full assessment. Knowledge of this for the carer and their family may help them to take up respite or feel more comfortable with the care they are providing, especially if they are subject to anger, irritability and problem behaviours – knowing it is has organic cause and not a response to the disease and its impact or the standard of their care.

The disease may have a great emotional toll on both carer and the person living with MND. There may be fear around increasing dependency and becoming a burden. There may be fear of the unknown, of the dying process and death, and a fear that if they are “too difficult” they may be abandoned or put into care. Anger arises in response to poor communication, function and mobility, changes in status and power, powerlessness and helplessness, and the reactions of others to their disabilities and condition. There are multiple ongoing losses and frustrations and everyday stresses that may result in anger.
In the face of multiple losses it is not easy to differentiate between sadness and depression. There is constant readjustment and reduced ability to enjoy daily activities. The prognosis can result in despair and lack of hope with respect to treatment and care. Treating depression can have a positive effect on the family. The effects of the responses to all of these issues impacts greatly on the family – especially the carer who may well have undiagnosed and unseen health issues of their own.

The need to integrate painful awareness with day-to-day living often means that people are able to interact with painful issues for brief periods of time but then the patient and family have the right to retreat from an unbearable truth and to process the pain and the reality as they are able. “Middle knowledge” is the concept of knowing and not knowing at the same time; a partial denial and partial awareness of death. This idea of middle knowledge captures the interaction and retreat around just what the patient understands about the finality of their prognosis (Weisman, 1972). This makes conversation about different options and health care issues problematic for the health professional, as they must ascertain just where the person is and what they understand in order to have a productive dialogue that is timely and sensitive (Weisman, 1972).

There are forced changes to roles and relationships, learning new skills at a time of distress and managing the impact of the disease and the severity and progression. Add to this distress and fatigue, changes to sexuality, intimacy, identity and financial and legal concerns, and the support of a palliative care agency seems essential (Oliver, 2008, p. 73). Carer burnout is a real threat to the continued care and management of the person with MND, so added support and assessment by a regular team of health professionals is important so that care needs can be carried out in a proactive, rather than reactive, manner.

**Health Professionals and MND Care**

Carers and health professionals must find ways to balance the needs of the person living with MND and other family members. It is helpful if isolation can be decreased and there is an awareness of the needs of other family members. MND Victoria has a range of support and information services that address these issues. It is important to create opportunities to express feelings without feeling guilty and to prevent carer burnout. It is helpful to address differing information and support needs.

Health professionals may be challenged caring for a person living with MND and they need to be in touch with their attitudes to issues such as disability, quality of life, euthanasia, and measures taken to prolong life. They may experience frustration with the inability of individuals to solve problems and subtle changes in patients’ cognitive ability may lead to difficulties in accepting advice or interventions. Health professionals will benefit from teamwork, some blurring of roles and communication via regular case conferencing. They will be working with feelings of helplessness and powerlessness and will benefit from self-care strategies, debriefing and supervision.

Early referral to palliative care enhances quality of life. It allows patient, carers and family to access a variety of specialist services, e.g. art and music therapy, volunteers, counselling, pastoral care and respite, as well as specialist care for comprehensive symptom management. Referral to palliative care facilitates top-up funding applications for community care and in-patient admissions, which provide for increased wellbeing, resources and care.

Referral can take place at any time postdiagnosis provided there is an unmet need that can be supplied by specialist palliative services. Early referral will allow conversations to be had that will establish a care plan based on the wishes of the person with MND, their carer and family, while they are able to communicate. They will receive practical and emotional support and education about death and the dying process as well as comfort and care within a holistic framework.

**References**


