# Palliative care in multiple sclerosis and motor neurone disease

***The general physician can help patients and families living with advanced multiple sclerosis and motor neurone disease in a number of practical ways. The key areas requiring a palliative approach are the management of physical symptoms, psychosocial support, decisions around potentially life-prolonging treatments and end-of-life care.***

Over the last 30 years palliative care has gone through many changes. It has evolved from caring solely for cancer patients and their families to helping people with non-malignant life-limiting conditions. People with multiple sclerosis and motor neurone disease may need palliative care to the same extent as people with cancer, experiencing uncontrolled pain, shortness of breath, fatigue and emotional distress (Newrick and Langton-Hewer, 1985; O'Brien et al, 1992; Higginson et al, 2006).

As well as coping with debilitating symptoms and progressive disability, these patients can be vulnerable to life-threatening episodes of infection and repeated hospital admissions. They may have to consider whether or not to have potentially life-prolonging interventions such as artificial feeding or respiratory support. These discussions can be difficult and frightening for patients and their families. Multiple sclerosis and motor neurone disease can place patients and families under significant levels of physical, emotional and financial strain, making multidisciplinary assessment and support essential.

It is not the case, however, that when active treatment stops, 'nothing more can be done.'

## When should we think of palliative care?

A person with multiple sclerosis or motor neurone disease may benefit from palliative care when there is an expected lifespan of 6–12 months, if there are distressing symptoms such as pain or breathlessness or if there is a need to consider end-of-life planning (*Table 1*).

A person dying from either disease should receive support from specialist palliative care services with regards to symptom control and end-of-life planning and families should be able to access bereavement support.

## Multiple sclerosis and motor neurone disease: different diseases, similar challenges

People living with both diseases display a similar range of needs encompassing physical symptoms and the possibility of psychological impairments, together with the emotional, social and financial impacts on themselves and their families. Multidisciplinary teams are well placed to support the patient and family.

An example of multidisciplinary teamwork in the context of multiple sclerosis and motor neurone disease is seen at St Catherine's Hospice in Scarborough. Monthly meetings are held to discuss the management of patients with team members consisting of a palliative care consultant, two neurology specialist nurses, the physiotherapist, occupational therapist, speech and language therapist and social worker.

Sudden and life-threatening deteriorations can feature in both conditions, commonly as a result of respiratory or urinary infections. Respiratory failure is the usual mode of death in motor neurone disease and can develop rapidly over hours. People living with multiple sclerosis and motor neurone disease often find it helpful to talk about the options of potentially life-prolonging interventions such as artificial feeding tubes and ventilatory support and understand more about the implications of having such treatments. Exploring peoples' preferences for future care is important and should be carried out sensitively and at a pace suited to the individual and family. As these people are at risk of developing communication and cognitive impairments, attempts should be made to maximize their capacity so that they can benefit from making informed decisions.

## Spectrum of symptoms in multiple sclerosis and motor neurone disease

Symptoms include physical, emotional and psychological domains.

### Multiple sclerosis

A survey of 50 people with advanced multiple sclerosis living at home and who were being referred to a specialist palliative care service revealed a high level of symptom prevalence (Higginson et al, 2006). Fatigue and spasms or pain were the commonest, with a reported prevalence of 70–80%. Swallowing problems and difficulty communicating affected 30–40%. Shortness of breath and

**Dr Julia Barnes** *is Specialist Registrar in Palliative Medicine and*
**Dr Colin Campbell** *is Consultant in Palliative Medicine, St Catherine's Hospice, Scarborough YO12 5RE*

*Correspondence to: Dr J Barnes*

nausea were troublesome for 25%. On average, these patients experienced nine symptoms each, varying from mild to severe. Levels of self-reported disability relating to mood and cognition were also graded moderate to severe.

The mean disease duration from diagnosis to referral was 18 years, with the range 0–55 years, reflecting the variability of disease progression.

### Motor neurone disease

A retrospective survey of 124 hospice patients with motor neurone disease also revealed a variety of symptoms (O'Brien et al, 1992). Speech and swallowing problems were the commonest with a prevalence of around 80%, and pain, cough, insomnia and shortness of breath affected around half of the people studied. Drooling of saliva was also common. In this group of patients with motor neurone disease, the mean survival from onset of symptoms until death was 42 months.

**Table 1. Spotlight on multiple sclerosis and motor neurone disease**

| | Multiple sclerosis | Motor neurone disease |
|---|---|---|
| Incidence | 3–7/100 000 | 2 /100 000 |
| Prevalence | 100/100 000 | 7/100 000 |
| No of cases in UK | 52 000–62 000 | 4000 |
| Typical age of onset | 20–50 years | 60–70 years |
| Disease trajectory | Protracted and unpredictable<br>Median survival 33 years from diagnosis<br>Primary progressive subtype (10%) associated with a much poorer prognosis<br>Around 50% of all sufferers develop secondary progressive multiple sclerosis which follows a degenerative course | Usually rapidly progressive<br>Average survival 2–3 years from diagnosis<br>Bulbar presentation associated with more rapid progression<br>Degree of involvement of respiratory muscles best predictor of life expectancy |

From National Institute for Clinical Excellence (2003); Department of Health Long-term Conditions National Service Framework Team (2005); Higginson et al (2006); Hirst et al (2008); McDermott and Shaw (2008)

## Pain in multiple sclerosis and motor neurone disease

Despite its high prevalence, pain is often under-recognized and under-treated. In multiple sclerosis, the severity of pain can rival that experienced by people with rheumatoid or osteoarthritis (Kalia and O'Connor, 2005). Pain becomes more common and severe as motor neurone disease progresses. It adversely affects quality of life in terms of physical functioning and mood, and contributes to a person's sense of suffering (Ganzini et al, 1999; Kalia and O'Connor, 2005) (*Table 2*).

Pain assessment involves a detailed history, examination findings and appropriate investigations as it is important to be able to distinguish between pain caused by the disease and pain caused by the disability. With the latter, it may be more appropriate to address issues such as wheelchairs, mobility aids, pressure area relief and constipation before changing the analgesia.

Neuropalliative rehabilitation describes the management of neurological patients who have significant disability, complex needs and a potentially shortened lifespan. It incorporates assessment and diagnosis of clinical problems, rehabilitation to maintain function, coordination of care and palliation of symptoms. Physiotherapists play a key role in optimizing the physical function of multiple sclerosis and motor neurone disease patients (*Figure 1*).

**Table 2. Types of pain in multiple sclerosis and motor neurone disease**

| Pain | Example | Mechanism | Therapeutic approach |
|---|---|---|---|
| Central neuropathic extremity pain in multiple sclerosis | Continuous extremity pain – may be burning in character, often worse at night | Multiple sclerosis plaques in spinal cord (pathological involvement of sensory pathways in motor neurone disease has been reported, but is rare) | Gabapentin titration according to efficacy and side effects<br>Amitriptyline can be added starting at a low dose – anticholinergic effects may limit dose<br>Opioids may be helpful |
| Central neuropathic intermittent pain in multiple sclerosis | Multiple sclerosis – trigeminal neuralgia – can be bilateral<br>Lhermitte's sign | Possible lesions at trigeminal nucleus or nerve root<br>Possible hypersensitivity of demyelinated cervical sensory axons | Carbamazepine usually used first line with phenytoin second line<br>Opioids may be helpful |
| Spasticity pain: seen in multiple sclerosis and motor neurone disease | Lower limbs may be more frequently affected.<br>Extensor and flexor spasms are seen in advanced multiple sclerosis | Multiple sclerosis plaques or motor neurone disease degeneration in the upper motor neurones | Baclofen – commonly used, starting with low doses and titrating upwards. It is better tolerated than diazepam, which can cause sedation and exacerbate weakness*. In severe cases intrathecal baclofen given continuously via a pump may be indicated<br>Physiotherapy and occupational therapy assessments |
| Musculoskeletal pain – common in motor neurone disease, especially if unable to turn unaided in bed | Back pain from prolonged sitting or lying<br>Muscle cramps<br>Skin pressure pain | Nociceptive pain arising from insult to muscle, tendon, ligament, soft tissue<br>Muscle wasting producing altered stresses on bones or joints | WHO analgesic ladder, but often benefit from modified release morphine starting at the smallest possible dose<br>Non-pharmacological: heat pads, TENS machine, massage.<br>Physiotherapy and occupational therapy assessments |

* from Beard et al (2003). TENS = transcutaneous electrical nerve stimulation; WHO = World Health Organization

## Are opioids safe to use in motor neurone disease?

Opioids are helpful for pain, dyspnoea and cough and should not be withheld because of fears of causing respiratory depression (Oliver, 1998). The initial dose should be low, e.g. Oramorph 2.5 mg 4-hourly, and should be titrated to the patient's symptoms with regular monitoring of effect. The 12-hour modified-release morphine preparation is useful for nocturnal pain and should be started at the smallest possible dose, e.g. 5 mg twice daily. Whenever a strong opioid is commenced, a laxative and anti-emetic should also be prescribed.

## Mood and psychological state

In multiple sclerosis, depression is thought to be three times more common than in the general population (Ghaffar and Feinstein, 2007); it is under-recognized and -treated and can result in suicidal ideation. Anxiety is also thought to be common. While a combination of antidepressants and psychotherapeutic treatment such as cognitive behavioural therapy may be appropriate for some, others may be less able to learn new coping strategies.

Major depression is less common in motor neurone disease and self-reported depressive symptoms include feelings of sadness or anhedonia (Ganzini et al, 1999).

People with both conditions experience multiple losses in terms of their independence, physical abilities and quality of their personal and social relationships (Edmonds et al, 2007). Day hospice services and support groups can enable patients and carers to socialize and derive support.

## Cognitive impairment in multiple sclerosis and motor neurone disease

In multiple sclerosis, nearly half of all patients may ultimately develop cognitive impairment and deficits are seen in concentration, executing tasks and short-term memory (Bobholz and Rao, 2003). People with motor neurone disease can also experience deficits in problem solving, attention and word generation (Massman et al 1996). Cognitive impairment affects a person's relationships, ability to interact and communicate with others and participate in discussions about care. Neuropsychological testing may be available and management strategies offered.

Cognitive impairment and communication difficulties can adversely affect a patient's mental capacity which has important consequences when it comes to considering and planning future care or life-prolonging interventions.

## Advance care planning

Palliative care teams and neurology teams have expertise in guiding patients in making future care decisions. These conversations should occur before swallowing and communication are lost and be introduced by a member of the health-care team who feels comfortable and competent in managing such conversations. There should be time for the patient and family to consider the implications of a life-prolonging treatment.

Some patients may not wish to engage in potentially upsetting and unpleasant conversations and this should be respected. However, while someone is able to communicate and has capacity there should be an exploration around his/her wishes.

A patient with capacity has the right to decline the offer of a medical intervention, even if that is perceived as an unwise decision by the medical team.

In all the following situations, where patients lack capacity, a valid advance decision relating to the withholding or withdrawal of life-prolonging treatment may apply or a lasting power of attorney may have been appointed. Where there is neither, a 'best interests' decision should be reached using multidisciplinary team working and in accordance with the Mental Capacity Act (British Medical Association, 2007; National Council for Palliative Care, 2008).

### Nutritional support

Choking or coughing when eating can be distressing and frightening to the patient and carer. Fatigue, difficulty in positioning the upper body, jaw spasticity and excessive saliva can all contribute. Addressing any potential treatable causes is important, for example ensuring good dental care and treating oral thrush. Anti-muscarinic medication can reduce saliva. Speech and language and dietetic assessments should be undertaken.

Patients at risk of losing their ability to swallow should be involved in discussions as to whether they would wish to be considered for artificial nutrition via a percutaneous endoscopic gastrostomy, or percutaneous radiological gastrostomy tube. The percutaneous radiological gastrostomy can be inserted without sedation, making it a safer option for people with motor neurone disease who have significant respiratory muscle weakness (usually defined as a forced vital capacity < 50% predicted). The likely benefits, risks and burdens of artificial nutrition should be explained.

#### Considerations in artificial nutrition

Benefits include the supplementation of food and drink, liquid medication can be given through the tube and it is a potentially life-prolonging intervention. Risks include those associated with fitting the tube, the chance of aspiration is not eliminated and the tube can become displaced.

Such an intervention carries with it possible burdens. An intervention that can prolong life will not be appropriate for imminently dying patients who should otherwise receive nursing and medical care to ensure comfort and a peaceful death. As part of the overall decision-making process consideration should be given to the circumstances under which artificial feeding would be stopped. As provision of artificial nutrition and hydration is a medical intervention, there may come a time

when the burdens of the intervention will outweigh the potential benefit and patients should be made aware that under such circumstances the feeding can be withdrawn.

Management of the feeding regimen and maintenance of the tube will require time and effort from the carer, whether family or a paid carer, and the primary care team.

### Management of infections where antibiotics may be withheld or withdrawn

Someone who is increasingly debilitated and experiencing frequent and severe episodes of chest or urine infections requiring admission to hospital for intravenous antibiotics may eventually want less aggressive management. It may be that optimizing quality of life and remaining at home for conservative management of infections will be the patient's and family's priority as the final stage of the illness draws nearer.

The patient may wish to be considered for admission to a hospice for treatment of infections, where further discussions can occur around the benefits and burdens of antibiotic therapy. Palliative care teams in the community and hospital can offer advice and support to the patient, family and treating teams in these situations.

### Shortness of breath in the patient with motor neurone disease

This is common and symptoms may include fatigue, daytime somnolence and morning headaches as a result of nocturnal hypoventilation. Characteristically it progressively worsens with disease progression, although acute deteriorations can occur, especially where there is significant bulbar impairment. It is a misconception that patients can 'choke' to death (O'Brien et al, 1992). The commoner modes of dying in motor neurone disease are either rapid decline over 12–24 hours from ventilatory failure, or progressive drowsiness secondary to hypercapnia.

Management should be multidisciplinary and include physiotherapy assessment to promote optimal positioning and breathing techniques. Associated anxiety can respond to a quick-acting benzodiazepine such as sublingual lorazepam. Opioids are also helpful for dyspnoea (Oliver, 1998).

Conversations about the possible benefits and burdens of non-invasive ventilation should be introduced at an early stage. Non-invasive ventilation can be used at home and in patients without significant bulbar involvement it can improve quality of life and survival for longer than riluzole (Bourke et al, 2006). In patients with bulbar involvement, quality of life improves but there is no survival advantage (Bourke et al, 2006).

## KEY POINTS

- Multiple sclerosis and motor neurone disease are progressive, life-limiting illnesses which may inflict many burdens and challenges upon the patient and family or carer.
- Physical symptoms are diverse and include pain, fatigue, breathlessness, plus speech and swallowing problems.
- Patients face multiple losses and adjustments which can have emotional and psychological consequences on themselves and those around them.
- The general physician is often the key professional ensuring that hospital patients with multiple sclerosis and motor neurone disease receive excellent palliative care, but health-care professionals must be able to recognize when these patients need specialist palliative care.
- Palliative care teams can help with difficult symptom control and end-of-life care, psychosocial support, plus decisions around potentially life-prolonging treatments.

## Goals of care in the last days of life

It is important, wherever possible, to recognize when someone with multiple sclerosis or motor neurone disease has entered the final stage of his/her illness. In multiple sclerosis, there can be repeated episodes of life-threatening sepsis, each of which could be the terminal event. In motor neurone disease, usually there is a progressive deterioration in symptoms but sudden deteriorations can occur and the patient (if appropriate) and family should be prepared for this. Good communication is essential to allay fears. The care pathway and place of death should be appropriate for the patient's needs, taking into account any previously expressed preferences.

Both in and out of hospital, the Liverpool Care Pathway incorporates anticipatory prescribing of subcutaneous key drugs for use as required. These may include:

- Diamorphine 2.5 mg (if opioid naïve) for pain
- Midazolam 2.5–5 mg for distress, breathlessness and agitation
- Hyoscine butylbromide 20 mg for distressing secretions
- An anti-emetic such as metoclopramide 10 mg.

A syringe driver enables continuous administration of essential medications.

The Liverpool Care Pathway also emphasizes the spiritual and psychosocial care of the dying patient and bereavement support for the family and carers.

## Conclusions

Particularly in the more advanced stages of multiple sclerosis and motor neurone disease, patients need and deserve all the diagnostic skills of the astute physician and the multidisciplinary team, coupled with a detailed knowledge of the therapeutic options. Palliative care in these diseases is grounded in meticulous attention to detail, thorough assessment and sensitive communication. **BJHM**

*Conflict of interest: none.*

Beard S, Hunn A, Wight J (2003) Treatment for spasticity and pain in MS – a systematic review. *Health Technology Assessment* **7**(40)

Bobholz J, Rao S (2003) Cognitive dysfunction in multiple sclerosis: a review of recent developments. *Curr Opin Neurol* **16:** 283–8

Bourke S, Tomlinson M, Williams T, Bullock R, Shaw P, Gibson G (2006) Effects of non-invasive ventilation on survival and quality of life in patients with amyotrophic lateral sclerosis: a randomised controlled trial. *Lancet Neurol* **5:** 140–7

British Medical Association (2007) *Withholding and Withdrawing Life-prolonging Medical Treatment: Guidance for decision making.* 3rd edn. Blackwell, Oxford

Department of Health Long-term Conditions National Service Framework Team (2005) *National Service Framework for Long Term Conditions.* Department of Health, London

Edmonds P, Vivat B, Burman R, Silber E, Higginson I (2007) Loss and change: experiences of people severely affected by multiple sclerosis. *Palliat Med* **21:** 101–7

Ganzini L, Johnston W, Hoffman W (1999) Correlates of suffering in amyotrophic lateral sclerosis. *Neurology* **52**(7): 1434–40

Ghaffar O, Feinstein A (2007) The neuropsychiatry of multiple sclerosis: a review of recent developments. *Curr Opin Psych* **20:** 278–85

Higginson I, Hart S, Silber E, Burman R, Edmonds P (2006) Symptom prevalence and severity in people severely affected by multiple sclerosis. *J Pall Care* **22**(3): 158–65

Hirst C, Swingler R, Compston D, Ben-Shlomo Y, Robertson N (2008) Survival and cause of death in multiple sclerosis: a prospective population-based study. *J Neurol Neurosurg Psychiatry* **79:** 1016–21

Kalia L, O'Connor P (2005) Severity of chronic pain and its relationship to quality of life in multiple sclerosis. *Multiple Sclerosis* **11:** 322–7

McDermott C, Shaw P (2008) Diagnosis and management of motor neurone disease. *BMJ* **336:** 658–62

Massman P, Sims J, Cooke N, Haverkamp L, Appel V, Appel S (1996) Prevalence and correlates of neuropsychological deficits in amyotrophic lateral sclerosis. *J Neurol Neurosurg Psychiatry* **61:** 450–5

National Council for Palliative Care (2008) *The Mental Capacity Act in Practice: Guidance for end of life care.* National Council for Palliative Care, London

National Institute for Clinical Excellence (2003) *Management of Multiple Sclerosis in primary and secondary care.* Clinical Guideline 8. National Institute for Clinical Excellence, London

Newrick P, Langton-Hewer R (1985) Pain in motor neurone disease. *J Neurol Neurosurg Psychiatry* **48:** 838–40

O'Brien T, Kelly M, Saunders C (1992) Motor neurone disease: a hospice perspective. *BMJ* **304:** 471–3

Oliver D (1998) Opioid medication in the palliative care of motor neurone disease. *Palliat Med* **12:** 113–15