What palliative care can do for motor neurone disease patients and their families

Appropriate palliative care is essential for people with motor neurone disease, a progressive and irreversible neurological condition that can have distressing symptoms in its later stages. **David Oliver** and **Samar Aoun** detail how and when the specialist palliative care team should get involved

Motor neurone disease (MND), or amyotrophic lateral sclerosis (ALS), is a progressive neurological condition with no curative treatment and a short prognosis – of two to five years from first presentation. The cause is unknown for the majority of patients, although in 5–10% of them there is a family history. Recent discoveries of new genes linked with MND suggest that there may be increasing evidence of a genetic cause, with an environmental stimulus leading to the disease in susceptible people.[1] The only treatment available is riluzole, which has been shown to slow the rate of progression – but probably only by a few months – over a period of two years.[2]

Key points

- Motor neurone disease (MND) is a progressive neurological condition with no cure and a short prognosis. Palliative care involvement is therefore appropriate from the time of diagnosis.
- The symptoms of MND, which are varied and will worsen as the disease progresses, should be managed appropriately.
- The psychosocial care needs of patients and their families or carers should be addressed, including when there are concerns regarding a possible genetic cause of the disease.
- The involvement of the specialist palliative care team may be episodic, occuring at times of change, crisis or decision-making; for example, at diagnosis, when discussing gastrostomy or ventilatory support, when there are cognitive changes and at the end of life.
- End-of-life care planning – and reassurance that, with good palliative care, death is usually peaceful – are a crucial part of care.

A palliative approach in MND

People with MND may present with variable and mixed symptoms and signs, but there are three main groups:

- People with ALS – with a mixed pattern of upper and lower motor neurone nerve damage, leading to weakness, wasting and increased reflexes
- People with progressive bulbar palsy – with primarily speech and swallowing issues
- People with progressive muscular atrophy – with predominantly lower motor neurone damage and increasing weakness and flaccidity of the legs.

However, as the disease progresses, these different patient groups may develop symptoms and signs in all body areas. Patients may rarely present with respiratory muscle weakness, leading to respiratory failure.

As there is no cure for MND, it can be argued that palliative care is appropriate from the time of diagnosis, especially as the average time between symptom onset and diagnosis is ten months (many patients are already severely disabled at diagnosis). Figure 1 shows the role of a palliative approach in MND from diagnosis to bereavement and the changes in care requirements as the disease progresses.[3]

In accordance with the WHO definition of palliative care,[4] the aims of care for people with MND should be to reduce the effects of the disease on the patient and their family; maintain the patient's abilities for as long as possible; and enable the patient and family to live as full a life as possible.

Holistic palliative care for MND patients and their families includes various aspects, which are outlined below. Symptoms need to

be carefully assessed and the psychosocial aspects need to be considered at all times.[5]

Symptom management

Symptom burden

MND patients will have many symptoms and these will vary from person to person. The burden will increase as the disease progresses,[5] the most common symptoms being: dysphagia (87% of patients), dyspnoea (85% of patients), weight loss (84% of patients), speech problems (74% of patients), pain (73% of patients), constipation (53% of patients), drooling (23% of patients) and emotional lability (23% of patients).[6]

Pain

Pain is common in MND and may be due to:

- Musculoskeletal pain – from changes around joints because of altered muscle tone, causing joint pain and/or stiffness
- Cramp – from increased muscle tone, especially following upper motor neurone damage
- Skin pressure pain – from reduced changes of position or immobility.

Careful positioning, physiotherapy and occupational therapy should all be considered. Specific medication – such as non-steroidal anti-inflammatory drugs for joint pain or antispasmodic medication for cramp – can be helpful. Many patients with skin pressure pain are helped by regular analgesia; they will start on simple analgesics, but many may require opioids.[7] [ref changed from 5 to 7; correct?] Studies have shown that morphine can be used both effectively and safely in MND.[8]

Dysphagia

As bulbar muscles weaken, swallowing can be affected, as there is reduced ability to chew food, move the bolus back within the mouth and swallow safely. Careful feeding, changing the consistency of food (to that of custard) and allowing time to eat are all helpful. However, many patients may need to consider gastrostomy – either percutaneous endoscopic gastrostomy (PEG) or percutaneous radiological gastrostomy (PRG).[9]

Careful discussion is necessary about the insertion of a gastrostomy. It will enable to continue nutrition and often improve quality of life, but there is little evidence that it will prolong life. The associated risks increase as respiratory function reduces, particularly with PEG insertion, where morbidity and mortality increase if the forced vital capacity is below 50% of the expected value. Discussion is therefore needed before swallowing is greatly affected and thus before the patient is really aware of the problems they are going to face.[10]

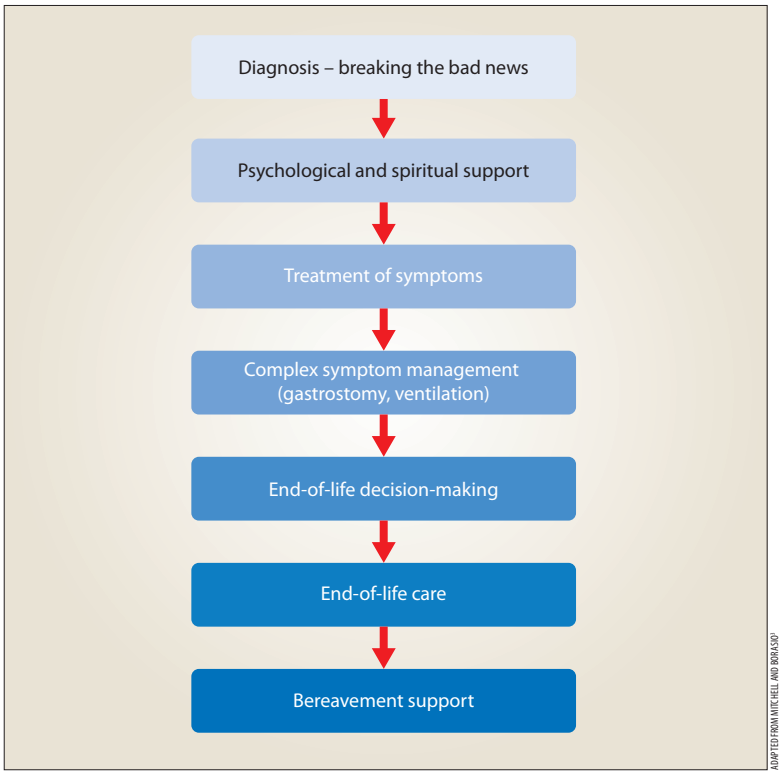


Figure 1. A palliative approach in motor neurone disease

Drooling

A significant problem associated with dysphagia is drooling; as the patient swallows less saliva than usual, it collects in the mouth and dribbles out. Good oral hygiene is crucial and some drugs may help: anticholinergic medication (such as sublingual or transdermal hyoscine), antidepressants (such as amitriptyline) or botulinum toxin injected into the salivary glands.[11]

Respiratory issues

As respiratory muscle and diaphragmatic function reduces, patients may become breathless or develop symptoms of respiratory failure (particularly at night). Symptoms will include orthopnoea, vivid dreams and nightmares, multiple arousals, morning headache, anorexia and generally feeling unwell. Careful assessment of respiratory function is essential in the ongoing care of the MND patient and respiratory support may need to be considered.[12]

In the UK, the National Institute for Health and Care Excellence (NICE) has produced

guidelines for the monitoring and assessment of respiratory function, and suggests considering non-invasive ventilation (NIV) in symptomatic patients.[13] Although there is evidence that NIV will extend life,[14] the disease will continue to progress, which entails the possibility of increasing disability, increasing dependency on NIV and decreasing quality of life. The NICE guidelines specify that, during assessment for and when starting NIV, end-of-life issues – including the withdrawal of NIV at the end of life – should be discussed.[13]

Tracheostomy ventilation may be an option for some patients, but there are even greater ethical issues than with NIV. The use of tracheostomy ventilation varies greatly, within Europe and globally.

Psychosocial aspects of care

Psychological support

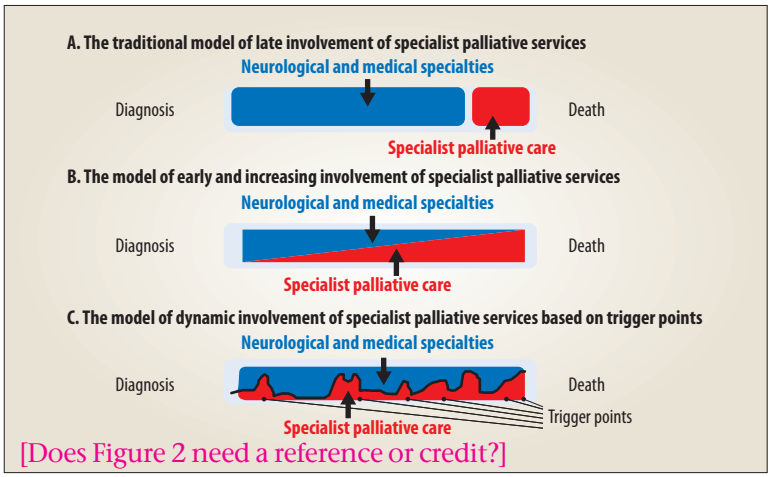
People with MND must cope with multiple losses – of their abilities, cognition, emotions – as well as face many difficult symptoms and increasing dependency.[15] They may have specific concerns about the diagnosis – the general media and the internet often emphasise the distress associated with MND and exacerbate the fear of choking to death. There may be particular issues for people with familial MND, as they may have memories of MND and deaths in their family and worry about their children. To allow these issues to be expressed, careful listening and support are crucial. Early involvement of palliative care will increase the chance of the patient still being able to express their concerns easily, before communication aids are needed.

Family concerns

The families and close carers of people with MND may have similar issues – experience of multiple losses, communication problems as the person's speech deteriorates – but also issues that are particular to them – their own fears and emotions, financial concerns.[16] Psychosocial support needs to be routinely offered to MND family carers throughout the illness trajectory. Family carers who get timely support from palliative care services will have better bereavement outcomes.[17]

Spiritual issues

The spiritual concerns of patients, families and carers (including 'why me?' and concerns and fears about death and dying) may deepen as the disease progresses. Together with cultural and religious issues, these concerns should be acknowledged and appropriate support should be provided.[18]



[Does Figure 2 need a reference or credit?]

Figure 2. Involvement of palliative care in motor neurone disease and other neurological diseases

A multidisciplinary approach

The multiple issues and concerns outlined above show that a multidisciplinary approach is required to managed MND. A team of appropriate health- and social care professionals is needed to assess and address all aspects of care. To ensure care is coordinated and patients and families feel supported, it is important that the team regularly works with MND patients, has the necessary experience and knowledge of the issues involved, and that its members work together. The wider caring team, including family members providing day-to-day personal care, also needs ongoing support, as the care of a person with MND can be physically and emotionally demanding.[16]

This team approach can be complex, as many different disciplines may become involved: neurology, rehabilitation, gastroenterology (if gastrostomy is being considered), respiratory (if NIV is used), as well as primary care and palliative care. There may be issues within and between the different teams involved, as these may have different attitudes and philosophies, and it is important that there is awareness of these issues.[19]

When is palliative care appropriate?

The provision of palliative care for people with MND varies, both between and within countries. In the UK, there is evidence that specialist palliative care services may be less involved in the early stages of the disease now than they were in the past.[20] However, there is increasing evidence that a wider

multidisciplinary team approach is effective, and may even extend patients' lives.[21]

Palliative care needs may vary through the disease trajectory and the involvement of specialist teams may be episodic, occuring at times of change, crisis or decision-making; for example, at diagnosis, when discussing gastrostomy or ventilatory support, when there are cognitive changes and at the end of life (see Figure 2).[22]

When more complex interventions are introduced, there may be new issues arising. For example, when gastrostomy or NIV are considered, there may be an emphasis on the discussion regarding the intervention and hence less discussion on how to manage the patient's progressive deterioration. This may in turn reduce everybody's awareness of the general deterioration and, crucially, of the need to discuss end-of-life issues. This has been addressed in the NICE guidelines, which say that end-of-life issues should be discussed when NIV is started and also when it is increasingly used as the patient approaches the end of life.[13]

Certain signs suggest that end-of-life care is needed, in patients with neurological disease in general (dysphagia, cognitive decline, repeated infections, weight loss, marked decline in condition) and in those with MND in particular (breathlessness/respiratory failure, swallowing issues, cognitive change, weakness).[23] These signs should act as triggers to encourage professionals to discuss end-of-life issues with patients and families and help them to start planning, looking at:

- Preferences for place of care and death
- Provision of anticipatory medications so that these are readily available if needed quickly (as suggested in the UK by the MND Association in its 'Just in case kit')[24]
- Advance care planning (advance directives, advance decision to refuse treatment, defining a proxy for decisions, and so on).

Conclusion

In any care setting, palliative care is essential for patients with MND and families to ensure their symptoms and issues are clearly identified and appropriately managed by a multidisciplinary team. Sensitive end-of-life discussions and planning will help as the end of life approaches. Many patients and families fear a distressing death accompanied by pain, choking and breathlessness. However, with good palliative care, death in MND patients is usually peaceful.[25,26] Reassuring patients and families is a crucial part of care.

Declaration of interest
The authors declare that there is no conflict of interest. [OK?]

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