

# Palliative care for patients with MND/ALS

**David Oliver** and **Gian Domenico Borasio** look at the role of palliative care in treating patients with motor neurone disease (amyotrophic lateral sclerosis)

**M**otor neurone disease (amyotrophic lateral sclerosis) is a relatively rare progressive, degenerative neurological disease with a prevalence of about 7 per 100,000 population.<sup>1</sup> The cause is usually unknown, although in about 5% there is a family history and of these in 20% (that is, 1% of the total MND/ALS population) an abnormal gene can be found – the superoxide dismutase 1 gene on chromosome 21.<sup>2</sup> There is no curative treatment but the glutamate blocker, riluzole, has been shown in trials to reduce the rate of progression and may extend life by a few months.<sup>3</sup> The prognosis is in the order of three to five years.<sup>4</sup>

In MND/ALS, there is progressive damage to motor neurones and thus muscle function is reduced. Every patient is different and the symptoms and signs will vary according to the nerves affected:

- Mixed upper and lower neurone damage with weakness and wasting of muscles, increased reflexes and fasciculations, presents as hand or leg weakness
- Lower motor neurone damage with increased weakness and flaccidity, often presenting as weak arms
- Damage to neurones in the brainstem, presenting as speech or swallowing problems.

As there is no cure, the care of patients can be considered to be palliative from the time of diagnosis. The multidisciplinary team approach to the care of the patient can be very helpful in ensuring that the symptoms are managed effectively and that the person with MND/ALS can remain as active as possible.

## At the time of diagnosis

As the symptoms of MND/ALS are often subtle and may be unrecognised for some time, patients may present with advanced disease and many symptoms. The diagnosis of MND/ALS is often a shock to both patient and family, and



may be unknown to them all, or may lead to particular fears and concerns. The recent interest and high profile court cases, regarding euthanasia and physician-assisted suicide, have increased public fear about MND/ALS due to discussion of its 'distress'. Information about MND/ALS and involvement of the wider team can be very helpful in allowing these fears to be addressed. It is important to encourage communication within the family, so that the concerns of all can be shared.<sup>5</sup>

There is a need to be open with the patient and family and early involvement of the speech and language therapist can allow the timely provision of appropriate communication aids, including computers

## Potential symptoms

### *Physical weakness*

As patients become weaker, the increased involvement of the physiotherapist and occupational therapist will allow mobility to be

## Key points

- Motor neurone disease (amyotrophic lateral sclerosis) is a relatively rare progressive, degenerative neurological disease, with a prevalence of about 7 per 100,000 population.
- As there is no cure, the care of patients and their families can be considered to be palliative from the time of diagnosis.

maintained as much as possible. Early discussion of aids, such as sticks, frames and wheelchairs, is helpful so that the patient and their family can adjust to these new losses of independence.

### ***Dysphagia***

As the disease progresses, up to 87% of people will develop difficulties in swallowing,<sup>6</sup> due to weakness of the muscles in the mouth, pharynx and oesophagus. Careful feeding is essential, and alteration of the consistency of foods to soft solids can be very helpful.<sup>7</sup> The insertion of a feeding tube should be considered early and insertion arranged before respiratory function has deteriorated too much, as there is increased risk if the forced vital capacity is less than 50% of that expected.<sup>8</sup> In advanced stages, insertion of a feeding tube, if warranted, can be safely performed in combination with non-invasive ventilation (NIV).<sup>9</sup>

When swallowing deteriorates, drooling may occur. Anticholinergic medication such as scopolamine (hyoscine orally or as a transdermal patch) or amitriptyline may be helpful. Injections of botulinum B toxin into the salivary glands may be considered for severe cases.

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### ***Speech problems***

Over 70% of people may develop problems with communication.<sup>6</sup> The early involvement of a speech and language therapist can allow the timely provision of appropriate communication aids.<sup>10</sup>

### ***Dyspnoea***

Dyspnoea is a problem faced by up to 85% of people with MND/ALS<sup>6</sup> and may be helped by opioids (starting at a dose of morphine 2.5–5 mg/4 hours). If anxiety is prominent, benzodiazepines may be started. Chronic hypoventilation may cause poor and disturbed sleep, anorexia, morning headache, nightmares and lethargy. NIV can be very helpful in relieving these symptoms, but careful discussion is needed before it is started.<sup>11</sup> As the respiratory muscles and diaphragm weaken further, the symptoms will recur and it is important to have made plans as to how these will be managed. If not, there is the risk of a sudden deterioration and invasive ventilation via tracheostomy being started without time for full consideration of the consequences, which include progressive weakness and dependency. In 15–20% of patients, the patient can become totally 'locked-in' with no form of communication. It is very

important to reassure the patients that, whenever they may decide to stop NIV, all necessary care and appropriate medication will be available to ensure a peaceful death.

### ***Pathological laughing/crying***

This symptom, which occurs in up to 50% of patients, can be socially distressing and responds well to treatment with amitriptyline or fluvoxamine.

### ***Pain***

Up to 73% of people with MND/ALS complain of pain,<sup>6</sup> even though the sensory nerves are unaffected. Pain needs to be assessed and appropriately treated:

- Musculoskeletal pain, due to atrophy and the altered tone around joints, may benefit from physiotherapy and non-steroidal anti-inflammatory medication
- Muscle spasm, from spastic muscles, may be helped by passive movements and muscle relaxants such as baclofen. Muscle cramps can be relieved by quinine sulphate
- Skin pressure pain can be relieved by the use of regular opioids, such as morphine.<sup>12</sup>

### ***Psychological aspects***

As the person with MND/ALS faces the progressive deterioration with the disease they may experience a wide range of emotions – fear of the disease, of deterioration, of dependency, of disability, and of dying and death. There is a need to be open with the patient and their family and to allow discussion of these issues, particularly while the person can still speak, as discussion with a communication aid can become increasingly difficult and distressing for all concerned.

### ***Social aspects***

Most people with MND/ALS are part of wider relationships or families, who may have similar concerns – of the disease, deterioration, finances, dying and death, and communication with children.<sup>13</sup> Sexuality is an important issue for many patients and partners though it is often unrecognised.<sup>14</sup> These concerns need to be discussed and a social worker or counsellor may be helpful in encouraging wider discussion of the more difficult issues.

### ***Spiritual aspects***

The issues regarding the deeper meaning of life often come to the fore when facing a progressive,

life-threatening disease.<sup>15</sup> Opportunities should be provided for the patient and their carers to look at the spiritual aspects of care – which may not necessarily be religious. Cultural aspects of care are also important and need to be addressed.

## The final stages

Many people with MND/ALS fear the final stages of the disease, and there are particular fears of choking, dyspnoea and pain. However, research has shown that, with good palliative care, over 90% of people die peacefully and choking is very rare.<sup>16,17</sup> Often the terminal stage occurs when the person develops a respiratory tract infection and the changes develop over only a few days; 48–72% of patients deteriorate rapidly and die within 24 hours.

It is important to anticipate any sudden deterioration. Discussion between the person with MND/ALS, their family and the professional carers is essential, so that all are aware of the plans if a deterioration occurs. Medication may be provided such as: morphine for pain and dyspnoea;<sup>12,17</sup> midazolam for relaxation and sedation; glycopyrronium bromide for chest secretions; and buccal lorazepam or midazolam to reduce distress while awaiting help.

In the UK, the Motor Neurone Disease Association has developed the Breathing Space Programme.<sup>6</sup> This provides leaflets for the person with MND/ALS, their family and healthcare professionals to encourage discussion of these issues at the end of life. There is also a box for medication, so that it can be stored easily in the person's house and is readily available for any healthcare professional to use if there is a sudden deterioration. The presence of the medication, together with the ongoing discussion and support of the professional carers, can be very reassuring for all involved.

The majority of people wish to remain at home and with good support, anticipation and the involvement of a co-ordinated team approach this is often possible.<sup>16,17</sup> A multidisciplinary team approach is essential and regular meetings to co-ordinate care are helpful. There is a need for ongoing support from the time of diagnosis but the team member involved most closely may vary over time.

## Bereavement

After the death, the family may need bereavement support. They have faced a gradual loss of the person with MND/ALS, as the disease

has progressed. There may be mixed emotions of relief and then guilt. Support can be helpful, allowing these emotions to be expressed.<sup>18</sup>

## Conclusion

During the disease progression of MND/ALS, the person and their family face loss of mobility, speech, swallowing, breathing, while for the majority of people the mind remains clear and aware. Palliative care has much to offer in allowing people with MND/ALS to retain the capabilities they do have and to make the most of the activities that remain possible for them. With good, multidisciplinary palliative care, they can die peacefully and in control of their lives.

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*David Oliver, Consultant in Palliative Medicine, Wisdom Hospice, Rochester and Honorary Senior Lecturer in Palliative Care, Kent Institute of Medicine and Health Sciences, University of Kent, UK; Gian Domenico Borasio, Professor and Chief Attending Physician, Interdisciplinary Center for Palliative Care and Department of Neurology, University Hospital Grosshadern and University of Munich, Germany*

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