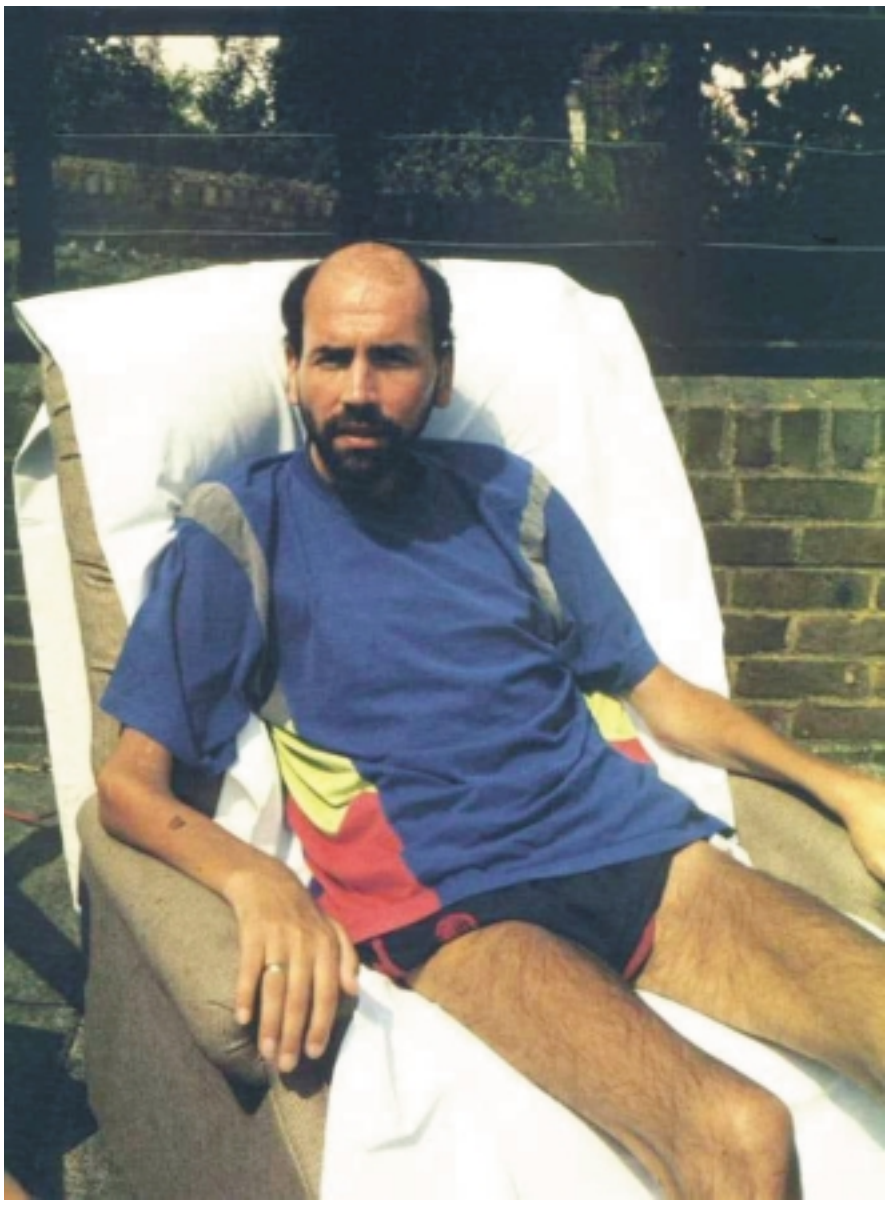

# Palliative care for motor neurone disease

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The palliative care of a patient with motor neurone disease (MND) starts even at the time of diagnosis, and this care throughout the disease process is crucial to management in the later stages.

Palliative care is defined as:

'The active total care of patients whose disease is not responsive to curative treatment. Control of pain, of other symptoms, and of psychological, social and spiritual problems is paramount. The goal of palliative care is achievement of the best possible quality of life for patients and their families.' (World Health Organization 1990).

As there is, at present, no cure for MND, and Riluzole, at best, only slows the progression of the disease. Therefore, the care of patients with MND is palliative and this approach will enable them, together with their families, to retain as good a quality of life as possible.

During the disease progression, there are times when there are particular care needs of the patient with MND:

- at diagnosis, when the patient and family are faced with a diagnosis that they may have little knowledge of and which may be very frightening;
- when a crisis in care occurs, such as the need to consider a new intervention, for example the use of a wheelchair, percutaneous endoscopic gastrostomy, ventilatory support;
- the terminal phase, when there is increasing deterioration and end of life issues are faced.

Often these phases are not easily defined. Indeed, different aspects of the progression of the disease may be faced at different times for different patients. All patients with MND are different, in the disease progression itself and how they, with their family and carers, cope with the many changes that they face. The challenge of palliative care of these patients is to ensure that *all* the various aspects of care are considered and actively managed, involving the patient and family in any decisions and changes. The aspects of care that need to be considered are:

- physical – symptom control, in particular;
- psychological – the effects on the patient;
- social – the effects on the family and carers;
- spiritual – concerns about the meaning of life and the challenges of the disease.

## PHYSICAL ASPECTS OF CARE

There are many different symptoms that should be considered, some are directly related to the disease process, others to the effects of muscle weakness.

### Dyspnoea

Weakness of the intercostal muscles and diaphragm can lead to increasing respiratory problems – in particular dyspnoea and respiratory failure. The patient may complain of the symptoms of respiratory failure, including morning headache, poor and disturbed sleep, dreaming, orthopnoea, mood disturbance and loss of appetite or of dyspnoea, sometimes on sitting up or when in a swimming pool (Lyall *et al*. 2000). These symptoms may be very subtle and careful assessment is essential, as patients may not realize they are developing respiratory problems. Dyspnoea is very common and up to 85% of patients develop the symptom during the disease progression (Oliver 1996).

A fuller assessment of respiratory function may be appropriate, including serial measurement of forced vital capacity, and tests of respiratory function, such as sniff analysis, blood gas estimation and overnight oximetry (Lyall *et al*. 2000).

Simple measures may be effective initially, such as ensuring the correct position with a slight incline backward in the wheelchair (O'Gorman & Oliver 1998) (Fig. 1.). The involvement of the wider multidisciplinary team is helpful, particularly the physiotherapist and occupational therapist (O'Gorman & Oliver 1998; Kingsnorth 2000). Opioid medication can be very effective if other methods of respiratory support are not appropriate or acceptable to the patient (Oliver 1998; Sykes 2000).

The palliative care of a patient with motor neurone disease starts even at the time of diagnosis

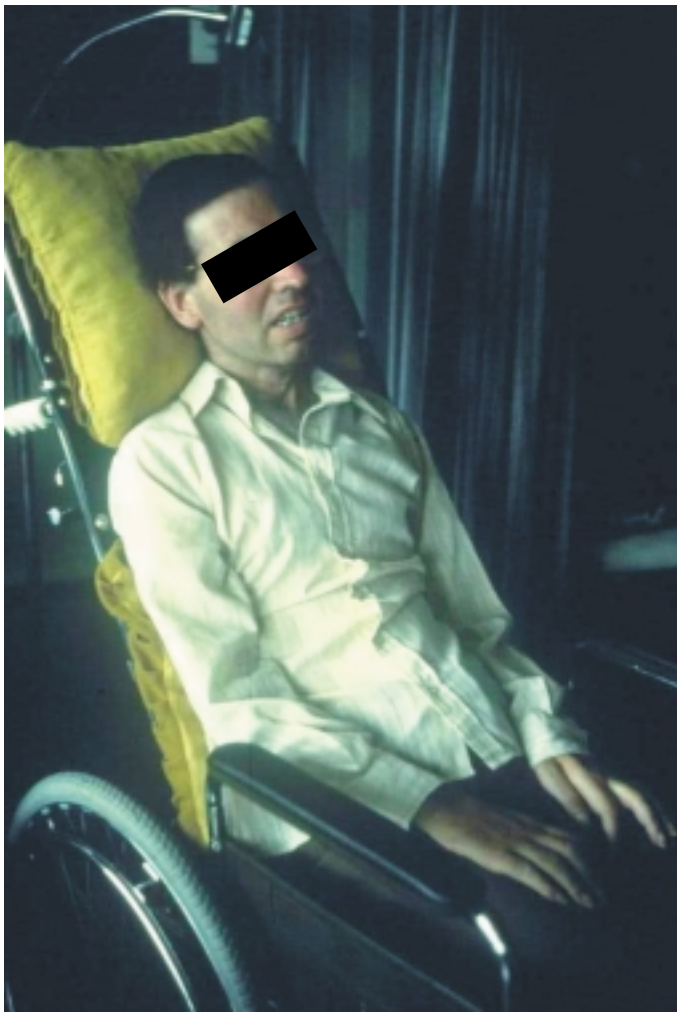

**Figure 1** A reclining wheelchair improves the comfort for the patient. Drooling was reduced and he no longer needed to hold a handkerchief to his mouth.

Consideration should be given to respiratory support, usually non-invasive positive pressure ventilation (NIPPV). NIPPV at night can be very helpful in reducing the symptoms of respiratory failure and so improving the quality of life. However, there must be full discussion with the patient and family because they should be aware of future plans and further management if the symptoms worsen again (Lyall *et al*. 2000). Many patients find that the use of NIPPV increases with time, until some require 24 h support. If there is further deterioration, there should be a clear management plan in this terminal phase.

On occasion, further respiratory support with tracheostomy ventilation may be considered. Unfortunately, this decision is often taken when there is a sudden deterioration, perhaps before the attending doctors realise that the patient has MND (Gelinas *et al*. 2000). There are risks that ventilation may put an increasing burden on the patients, and particularly their families (Gelinas *et al*. 1998), as they become increasingly disabled and need total care. Approximately 10% of patients become 'locked in', with no method of communication, but with the basic support of life continuing (Gelinas *et al*. 2000). Patients need to know about the risks of this development when NIPPV is discussed, so that they may take a clear and autonomous decision (Gelinas 2000). The use of ventilatory support opens up many areas of ethical debate and dilemmas, and all involved in the patient's care should be aware of the potential risks and the need to discuss these honestly.

## Dysphagia

The development of dysphagia is a distressing and common symptom, affecting up to 87% of patients suffering from the disease (Oliver 1996). A careful assessment is essential, and involves a speech and language therapist early in the disease progression (Wagner-Sonntag *et al*. 2000; Scott & Foulsum 2000). Careful feeding and the use of pureed food, to a consistency that eases swallowing for the individual patient, is helpful (Wagner-Sonntag *et al*. 2000; Scott & Foulsum 2000). The placement of a percutaneous endoscopic gastrostomy (PEG) tube can be very helpful in improving quality of life (Gazis *et al*. 1996) and there is evidence that it may increase survival (Mazzini *et al*. 1995). (Fig. 2.)

The placement of a PEG tube should be discussed early in the development of swallowing difficulties. The PEG tube must be inserted while respiratory function is relatively good – forced vital capacity of at least 50% of expected – to minimise morbidity and mortality from the procedure (Miller *et al*. 1999). Careful discussion with the patient and family is important so that they are aware of the potential benefits of PEG, and the PEG tube may not even be used for any feeding initially. However, as swallowing deteriorates, the PEG can be used increasingly, in conjunction with a decreasing oral intake (Wagner-Sonntag *et al*. 2000).

As a result of reduced swallowing, many patients develop drooling: 23% in one study (Oliver 1996). This can be very distressing for both patient and family, especially as on many occasions the patient may be treated by others as if they are of low intelligence. Anticholinergic medication can be effective in reducing saliva and drooling: atropine 300–600 □g tds; sublingual hyoscine 300 □g tds; hyoscine hydrobromide transdermal patch 1 mg/72 h (Scopoderm TTS), although this may cause

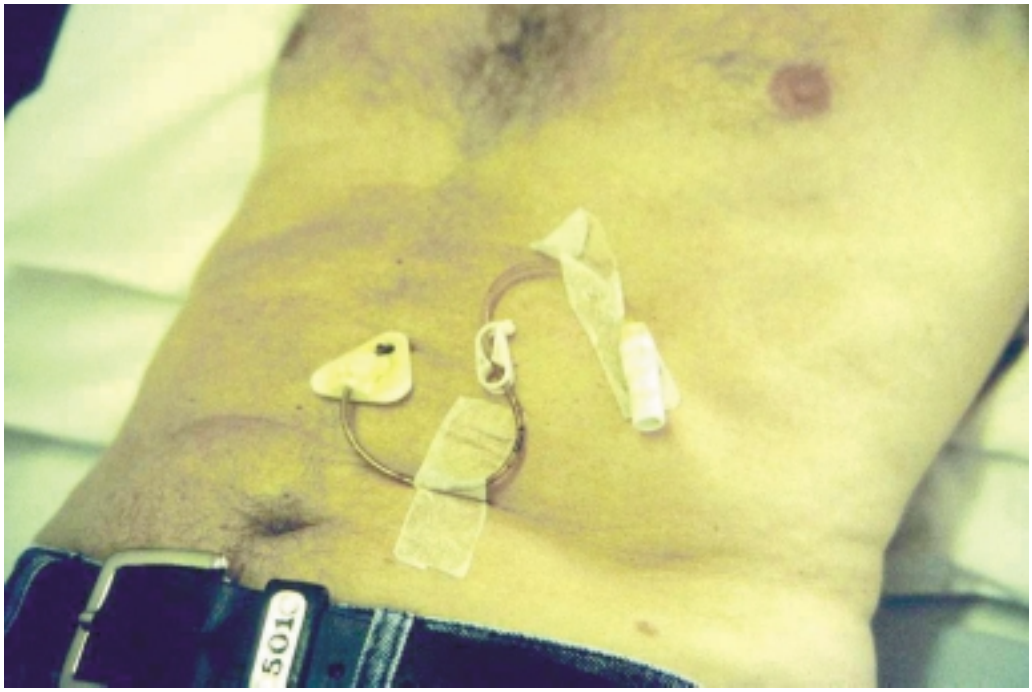

**Figure 2** The insertion of a PEG can improve the quality of life. This patient was able to visit his brother in Australia after the procedure, without the anxiety of whether he would be able to eat.

skin irritation. Tricyclic antidepressants – amitriptyline starting at 25 mg at night and slowly increasing – can also be helpful (Scott *et al.* 2000). Injection of Botulinum A Toxin into the salivary glands has been suggested (Geiss *et al.* 2000) although further research is required. On occasions, a continuous subcutaneous infusion by syringe driver of an anticholinergic drug may be required: glycopyrronium bromide 600 □g–1.2 mg/24 h; or hyoscine hydrobromide 600 □g–2.4 mg/24 h (Oliver 1996; Sykes *et al.* 2000). Radiotherapy to the salivary glands has been suggested but can leave a very dry mouth, or the production of thick secretions.

Some patients have great problems with thick tenacious secretions, which may be difficult to clear with poor respiratory muscle function. There is evidence that beta adrenergic receptors are involved in the production of these thicker secretions and propranolol – at an initial dose of 10 mg tds – has been found to be useful in limited trials (Sykes 2000; Scott & Foulsum 2000).

### Emotional lability

Emotional lability (pseudobulbar affect) is common in MND patients, particularly those with bulbar disease, and one study showed a prevalence of 23% (Oliver 1996). There is increasing evidence that this symptom is related to frontal lobe damage and dysfunction. It can be particularly distressing to both patients and families because the patient may feel as if their emotions are out of control and that they are going mad (Borasio & Voltz 1997). If there is a suspicion of emotional lability, especially in patients with bulbar disease, it is often helpful to explain that the symptom may occur. Patients may be relieved that the symptom is understood and that they can discuss their fears and concerns (Borasio & Oliver 2000). The explanation that the symptom is a known part of the disease and is understood may be the most helpful intervention in reducing anxiety.

Treatment with antidepressants, such as amitriptyline starting at 25 mg at night, can be helpful. Anxiolytics, such as benzodiazepines may also be helpful (Borasio & Oliver 2000).

### Weakness

Progressive weakness is inevitable as the disease progresses. There are no proven treatment regimes, and although creatine has been suggested to be of help, studies have yet to confirm this. Explanation is essential in allowing the patient and family to cope with the increasing weakness so they feel able to accept help, in the form of aids such as a stick, wheelchair or electric raising chair. Physiotherapy is very helpful in allowing the patient to make the most of their remaining power, and in adjusting to increasing disability (O'Gorman 2000).

### Pain

Although there is little evidence of sensory

nerve involvement, pain is a common symptom, with studies showing a prevalence of up to 73% (Oliver 1996). The pain may be due to:

- muscle stiffness and spasm in spasticity, so the use of muscle relaxants such as baclofen can be helpful (Borasio & Oliver 2000);
- joint pains due to the altered muscle activity around the joint – the physiotherapist can help in the assessment and management of this type of pain by ensuring good positioning and encouraging passive movements (O'Gorman 2000);
- skin pressure pain due to immobility

The treatment of skin pressure pain is initially with a simple analgesic, such as paracetamol or codydramol, on a regular basis. However, many patients benefit from the use of opioid analagesics. Regular morphine, as morphine elixir, starting at 5 mg every 4 h, or modified release tablets or capsules (MST or Zomorph) starting at 10 mg every 12 h, can be very effective (Oliver 1998). Studies have shown that morphine can be used safely if it is titrated carefully to the patient's pain and the median dose in one study was 60 mg/24 h and the mean duration of use was 95 days (Oliver 1998). Quality of life can be greatly improved, without necessarily shortening life, with the correct use of morphine. If swallowing is affected, a transdermal preparation, such as fentanyl transdermal patch, starting at 25 □g/ h, or a continuous subcutaneous infusion by syringe driver may be used at a dose of morphine or diamorphine equivalent to that given orally, or starting with (dia)morphine 15 mg/24 h (Sykes 2000). Diamorphine has no particular advantage over morphine, other than it is more soluble, and morphine can be used as effectively.

### Insomnia

From early in the disease, patients may find sleep difficult. A careful assessment is essential as there may be several causes:

- anxiety about the diagnosis, or disease progression;
- depression as a result of the disease process;
- fear of not being heard by carers when there is reduced speech, particularly if extra help is required in moving in bed;
- muscle cramps or spasticity;
- dysphagia and problems with clearing secretions, or fear of drooling;
- respiratory insufficiency, with nightmares, dreams and dyspnoea.

Causative factors must be sorted out before treatment is started. There may only need to be some simple action, such as the provision of a sensitive buzzer to allow a patient with reduced speech to call for help, or appropriate medication may be needed, such as antidepressants, muscle relaxants, anticholinergics or anxilolytics.

### Cough

Cough may develop:

- as the respiratory muscles weaken and it becomes increasingly difficult to clear secretions from the chest – the reduction of these secretions, particularly at night, may be helpful, in conjunction with physiotherapy;
- as swallowing efficiency declines and saliva trickles down into the airways, stimulating the cough reflex;
- if there is a respiratory tract infection, upper or lower, with increased secretions and a reduced ability to clear them.

Treatment should be of the underlying cause (Lyall *et al*. 2000). Opioids may be helpful if cough remains troublesome (Oliver 1998).

### Constipation

Constipation may occur in patients taking a reduced diet, or with reduced physical activity, particularly when medication has been prescribed that can lead to constipation, such as opioids or anticholinergics – one study reported 53% of patients complained of constipation (Oliver 1996). Prevention of constipation is important, by ensuring adequate hydration, a balanced diet and aperients (Borasio & Oliver 2000).

## PSYCHOLOGICAL ASPECTS OF CARE

From the time of diagnosis, and often before the diagnosis has been made, patients may have concerns and fears – of the diagnosis, the treatment, the disease progression, dying and death (Borasio *et al*. 2000). All those involved in the care of people with MND should be aware of these possible concerns and fears and be willing to explore these issues if and when necessary.

### Fears of the diagnosis

Many patients and their families have little prior knowledge of MND and fear the unknown. Unfortunately, many books emphasise the distress of the disease and death from MND. Moreover, the press coverage of people with MND has tended to stress the problems of the disease,

especially when covering the people with MND who have been in the courts pressing for the right to die. These issues may need to be discussed and the alternative views presented.

### Fears of dependency and disability

Many patients fear loss of control and the loss of independence as the disease progresses. Ways of coping with changes may need to be explored and psychosocial support, together with practical support, such as the provision of a wheelchair, may be necessary (Kingsnorth 2000; O'Gorman 2000).

### Fears of dying

These are common, especially fears of dying from choking or suffocating. There is now evidence that, with good palliative care, death is not distressing and most often it is peaceful (Neudert *et al*. 2001).

## SOCIAL ASPECTS OF CARE

Most patients are part of a wider group of family or carers. They too will be affected by the progression of the disease and may have similar concerns.

### Fears of the diagnosis

Similar to the fears of the patient, but there may be differences and it may be necessary to enable them to discuss their fears together. The support of a social worker or psychologist may be helpful (Borasio *et al*. 2000; Gallagher & Monroe 2000).

### Communication issues

These may occur within the family group, particularly when the diagnosis or prognosis are not shared within the family.

### Financial problems

These may occur when the patient and then their spouse or carer have to leave paid employment due to increasing disability or care needs. This can lead to stresses within relationships and increasing financial pressures. A social worker can be helpful in ensuring the necessary support is available (Gallagher & Monroe 2000).

### Sexuality

This may become an issue for many patients, although they may find this difficult to admit. The sexual practices of a couple may have to change as disability increases and they may need encouragement and help to discuss the issues together, and to consider alternatives. The need to express love is important to many couples and these issues, if not addressed, may lead to distress, which is often not acknowledged.

### Fear of dying

Families and carers may have their own fears about the process of dying: everyone has their own experiences and fears about death. They may need to be encouraged to express these concerns openly so they can feel more able to care for the patient and cope

Quality of life can be greatly improved, without necessarily shortening life, with the correct use of morphine

with both their own and the patient's concerns (Gallagher & Monroe 2000).

## SPIRITUAL ASPECTS OF CARE

As someone faces increasing disability and the possibility of an early death, they may start to consider the deeper meaning of life and death. This expression may not necessarily be in the form of organised religion, but in terms of 'natural religion', with concerns over the future. Time may be needed to talk about these issues and it is easier to do so while speech is reasonably intact, as to discuss these deep issues by means of an electronic communication aid can be very difficult.

## MULTIDISCIPLINARY CARE

To enable a full assessment of the problems and concerns of a patient with MND, and their family, it is essential that a multidisciplinary approach is used. The team includes:

- neurologist/physician;
- nurse;
- chaplain/religious leader;
- dietician;
- occupational therapist;
- physiotherapist;
- social worker/counsellor;
- speech and language therapist.

All these people may need to be involved in the care and support of the patient with MND and it is essential that there is a co-ordinated approach to the care provided. This may also involve other professionals and support workers, including:

- social services care manager, or link worker
- disability services;
- specialist nurses in neurology clinics;
- research workers in MND;
- Motor Neurone Disease Association: the Regional Care Advisors who are able to provide support and information for patients, families and professionals;
- the local MND Association Branch, with local volunteer visitors for support and the opportunity to meet and discuss MND care (Fig. 3);
- primary health care team of general prac-

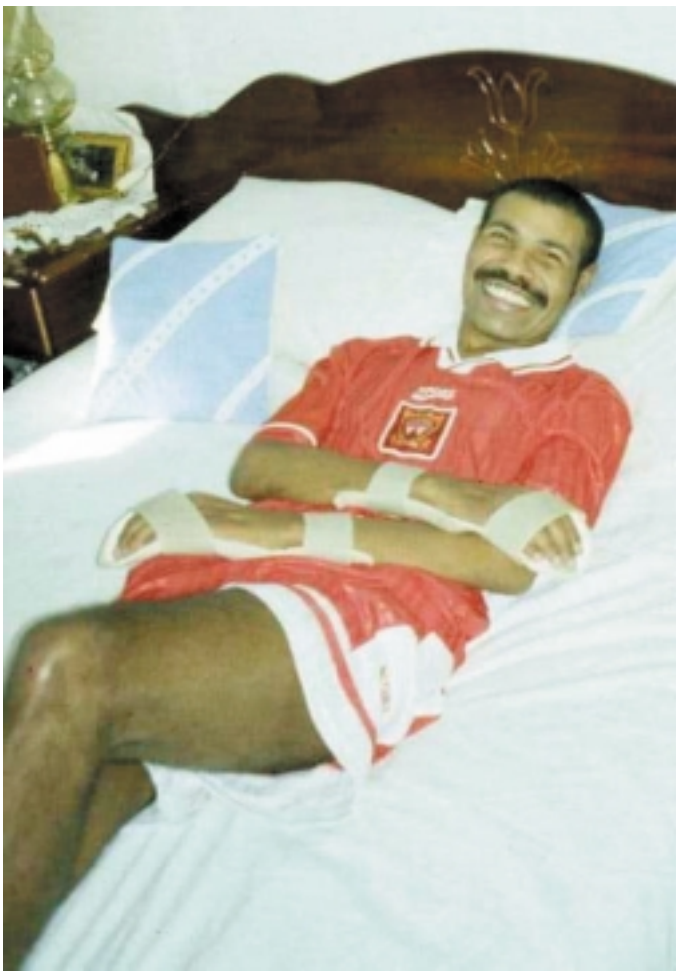

**Figure 3** Consideration of patient autonomy is important. This man in South Africa was able to remain at home with the support of his wife and family, and the assistance of the MND Association.

**Figure 4** Laptop computers and other communication aids can help communication and enable independence. This lady was able to continue communication with her family and carers, using the voice system.

titioner and community nurse who will be providing on going care at home.

With so many different people involved there is a real possibility of overlapping care, or missing areas of care. Moreover, the patient and family can feel overwhelmed, and confused as to what is being provided. Their own home may feel invaded by all the visiting professionals (Pegg 1994).

To facilitate multidisciplinary care, a co-ordinated approach is required. In many areas there are developing MND teams, meeting to discuss patient care, ensure a multidisciplinary approach and help in the education of other professionals (Holmes 2000). These may develop from different backgrounds, depending on the local expertise, including:

- neurology services and the MND care and research centres (Holmes 2000);
- rehabilitation services (Molloy 2000);
- palliative care services (Oliver 1996; Oliver 2000);
- the MND Association (Holmes 2000).

The aims of these teams are to:

- ensure a multidisciplinary assessment;
- co-ordinate the visits of professionals;
- provide a link for patients and families with the team, often via a link worker;
- co-ordinate the inter-agency working that is necessary;
- provide appropriate, and timely, information for patients and families, with sensitivity and respect;
- anticipate the emotional and physical needs of patients;
- offer support to patients and families, and to provide the information for autonomous decision making;
- ensure continuity of care all through the disease progression (Holmes 2000).

Throughout the team approach, it is essential that the team aims to facilitate and enable patients and families, focuses on their needs and concerns, and involves all the family in the care and decision making, with the patient's agreement.

The needs of patients and families will vary greatly according to the disease progression, the family's own resources and previous coping mechanisms, and the availability of resources and support. However, there may be specific needs as the disease progresses. At the time of diagnosis, there may be a particular requirement to provide information, and support patient and family with their concerns and fears. The way this information is imparted and the support provided at this time can affect the later care of the patient and family. Later reactions to problems can be profoundly altered by the care at this time. A supportive and empathetic approach, with the opportunity for questions and a sympathetic reaction by the professional, is very important in showing that support will be available later so that the patient and family know they can discuss issues openly with the caring team (Borasio *et al.* 2000; Gallagher and Monroe 2000). (Fig. 4.)

To anticipate a crisis it is often helpful to discuss the use of medication, and to provide the medication for the patient's house

As the disease progresses there may be specific times when members of the team should be involved, as problems evolve. As mobility decreases the physiotherapist and occupational therapist may be of most help. If speech and swallowing deteriorate, the speech and language therapist and dietician should be involved, and as early as possible so a relationship can be developed. With all these changes, there may be increasing needs for support and for a social worker, counsellor or clinical psychologist to be available to allow patients to talk about their fears and reactions (Kerkvliet 2000). Patients must be psychologically able to discuss new treatment plans or interventions, for if they are unable to discuss the issues the intervention may be rejected and the care offered compromised. For instance, the discussion about the insertion of a PEG tube may have to be undertaken over a period of time, allowing the patient to understand the problems and to face the issues involved. With the appropriate and sensitive provision of information, an autonomous decision can be taken more easily.

## THE TERMINAL STAGES

As the patient deteriorates, there is an increasing need to assess and look at all the concerns and problems of the carers and family as much as those of the patient. The control of symptoms is essential and the wider multidisciplinary team should be involved, in a co-ordinated way. The deterioration of a patient with MND is variable. Patients usually deteriorate slowly and it gradually becomes apparent that the disease has reached the terminal stage, but for many patients the changes are rapid, over a few days (Neudert *et al*. 2001). One study showed that for 65% of patients the period between the acute deterioration and death was under 24 h (Neudert *et al*. 2001).

The cause of death is usually respiratory failure, if invasive ventilation is not considered to be appropriate. The patient develops a respiratory tract infection, which initially may seem trivial, but the respiratory function deteriorates and over a few days, sometimes over a few hours, there is a rapid deterioration leading to death (Neudert *et al*. 2001). One study showed that 86% died from respiratory failure, 6% from heart failure, 4% from pneumonia, 1% from suicide and 3% from other causes (Neudert *et al*. 2001).

Patients and their families fear the deterioration, and in particular choking or suffocating (Oliver 2000; O'Brien *et al*. 1992). However, distress is rare and with good palliative care, including the management of dyspnoea, pain and swallowing problems, and support of patient and family, the vast majority of deaths from MND can be peaceful (Neudert *et al*. 2001; O'Brien *et al*. 1992). An international study showed that only 5% of a group of 171 patients in the UK and Germany died in distress, and 91% died peacefully (Neudert *et al*. 2001).

### Co-ordinated care

The care in the later stages of MND will involve carefully co-ordinated care. Anticipating crises is important – ensuring that all involved in the care are aware of the plans for care, so that if there

are increasing respiratory problems the patient, family and carers are all well aware of the patient's wishes so that unwanted and inappropriate intervention, such as ventilation, is avoided. The provision of medication for a crisis, together with discussion within the family, can be very helpful in allaying the concerns about a crisis for both patient and family. The medication is readily available if the crisis, such as breathlessness or pain, occurs out of hours and all are prepared for the actions to be taken in such an event (Oliver 1996; Sykes 2000) – see below. Symptom control is maximised, by reassessment on a regular basis and close involvement of the multidisciplinary team. Pain, dyspnoea and drooling should be addressed in particular. Opioids may be very helpful, as outlined above. As the patient deteriorates, oral medication may be difficult and alternatives have to be considered, such as a continuous subcutaneous infusion by syringe driver (Sykes 2000). This can allow opioids to be given, usually as (dia)morphine (starting at a dose appropriate to oral morphine, or at 10–15 mg/24 h), together with midazolam as sedation and to reduce stiffness (at a dose of 20–60 mg/24 h) and an anticholinergic to reduce respiratory secretions and respiratory distress (such as glycopyrronium bromide 1.2 mg/24 h or hyoscine hydrobromide 1.2–3.6 mg/24 h) (Oliver 1996; Oliver 1998; Sykes 2000).

To anticipate a crisis it is often helpful to discuss the use of medication, and to provide the medication for the patient's house. The 'Breathing Space' programme of the MND Association has been produced to facilitate this discussion about the dying process and there are leaflets for the patient and family and for the general practitioner with points for discussion. The MND Association also provides a box for the storage of medication, together with a leaflet on the suggested medication and its use (Fig. 5). The box has two parts:

- one section for professionals with the suggested medication [(dia)morphine, in a dose appropriate to the oral opioid, or at 5 mg–10 mg; midazolam for sedation and muscle relaxant at a dose of 5–10 mg; glycopyrronium bromide 200 micrograms or hyoscine hydrobromide at a dose of 400 micrograms for the reduction of respiratory secretions]; water for injections; syringes and needles;
- one section for families, if appropriate after discussion with the family, containing diazepam enema 10 mg to be given rectally by the family in the crisis situation while they are waiting for professional assistance (Oliver 1996; Sykes 2000).

The support of patient and family is very important at this time and regular contact is helpful. The family need to have a point of contact for advice at all times.


**Figure 5** The 'Breathing Space' programme. This allows the discussion of end-of-life issues between the patient and family and carers, and the provision of medication in anticipation of the development of a crisis.

The professionals involved in the care of people with MND may also benefit from support, as the patient deteriorates (Kerkvliet 2000). All too often the professional carers are ignored although they may also be affected by the deterioration and death of a patient who they have known for a long time. The multidisciplinary team can be very supportive if all are aware of these needs and a team meeting can help in providing this support (Kerkvliet 2000).

Communication among all involved is essential. It is important that all the team, including the primary health care team, are aware of the change in emphasis in care as the terminal stages of the disease are reached. Some members of the team may have an altered role in these later stages, for example occupational therapy requirements may reduce and the role of the physiotherapist is more focussed more on ensuring comfort than on aiding mobility. It is also important to ensure that the patient and family are aware of this change in care, and are fully involved in making these decisions.

### Specialist palliative care

Specialist palliative care can be very helpful throughout the disease progression and over 70% of hospices and other specialist palliative care providers in the UK are involved in the care of people with MND (Oliver & Webb 2000). The services can provide:

- support at home with the multidisciplinary home care (Macmillan) team providing support and advice for patient, family and primary health care team;
- support in the hospice or palliative care unit: respite care; symptom control and multidisciplinary team assessment; terminal care in the later stages of the disease;
- support for care at home with day hospice care, providing respite for families and the opportunity for assessment and socialization;
- support in hospital with hospital palliative care teams, providing support and advice for hospital staff;
- psychosocial and bereavement support (Gallagher and Monroe 2000; Oliver & Webb 2000).

In the past, the role of specialist palliative care has often been more in the later stages of the disease (O'Brien *et al*. 1992). However, for the provision of optimum care, this role is increasingly seen to be important from the time of the diagnosis, or soon after. In this way the specialist palliative care team can work together with other agencies and build up a relationship with the patient and family. This is particularly important when there are bulbar symptoms, because it is very difficult to help patients communicate their fears and deeper concerns if speech has already been lost and communication limited to a communication aid. If the aim of care is to enable the patient to be fully involved in their care, communication is important and the longer term relationship can facilitate this.

## BEREAVEMENT

The care of the family of someone with MND may continue after the death of the patient. Bereavement care is important, although for many families there have been so many continual losses throughout the disease progression that the death is only one further final loss (Centers 2000). There may be mixed emotions, with feelings of relief that the disease has ended, along with feelings of guilt that with such a loss relief is inappropriate. Counselling and support may be helpful for many families (Gallagher & Monroe 2000). Families may also ask for further advice and information about the risks of MND in the family and time may be needed to reassure them that their family is at no higher risk. In the rare families with familial MND, counselling and genetic testing may be necessary.

## CONCLUSIONS

The development of the Practice Parameters in the USA (Miller *et al*. 1999) has stimulated all involved in the care of patients with MND to look carefully at the care provided. Although, at present, there is little evidence for many of the interventions and treatments used, there is an increasing evidence base. The use of such guidelines will increase over the coming years (Mitchell 2000) and in the UK there are shortened guidelines for the management of MND (MND Advisory Group 1999).

The palliative care of MND starts at the time of diagnosis. With careful multidisciplinary assessment of the various concerns and problems of the patient and family, it is possible to reduce the effects of the disease progression, maintain the patient's remaining activities, and enable the patient and family to live life as fully as possible. We can now tell patients, that with good palliative care, quality of life can be maintained, and that death will usually be peaceful.

## USEFUL WEBSITES

- World Federation of Neurology site, with regularly updated news (www.wfnals.org);
- The International Alliance of ALS/MND Associations (www.alsmndalliance.org);
- The UK Motor Neurone Disease Association (www.mndassociation.org);
- The American ALS Association (www.alsa.org);
- The Scottish Motor Neurone Disease Association (www.scotmnd.org.uk);
- The Cananadian ALS Association (www.als.ca);
- The Les Turner Foundation in Chicago (www.lesturnerals.org);
- The American Muscular Dystrophy Association, involved in ALS care (www.mdausa.org).

## REFERENCES


Borasio GD & Voltz R (1997) Palliative care in amyotrophic lateral sclerosis. *Journal of Neurology,* **244** (Suppl. 4), S11–S17.

Borasio GD & Oliver D (2000) The control of other symptoms. In: Oliver D, Borasio GD Walsh D, eds. *Palliative Care in Amyotrophic Lateral Sclerosis*. Oxford, Oxford University Press, pp. 72–81.

Borasio GD, Sloan R & Pongratz D (2000) Breaking the news. In: Oliver D, Borasio GD Walsh D, eds. *Palliative Care in Amyotrophic Lateral Sclerosis*. Oxford, Oxford University Press, pp. 29–41.

Centers L (2000) One day at a time: the experiences of an ALS caregiver. In: Oliver D, Borasio GD Walsh D, eds. *Palliative Care in Amyotrophic Lateral Sclerosis*. Oxford, Oxford University Press, pp. 183–7.

Gallagher D & Monroe B (2000) Psychosocial care. In: Oliver D, Borasio GD Walsh D, eds. *Palliative Care in Amyotrophic Lateral Sclerosis*. Oxford, Oxford University Press, pp. 83–103.

Gazis A, Rawlings JR, Allison SP & Jefferson D (1996) A prospective study of nutritional changes and of gastrostomy feeding in motor neurone disease. *Proceedings of the Nutrition Society*, **55**, 146a.

Geiss R, Naumann M, Werner E *et al.* (2000) Injections of botulinum toxin A into the salivary glands improve sialorrhoea in amyotrophic lateral sclerosis. *Journal of Neurology, Neurosurgery and Psychiatry*, **69**, 121–3.

Gelinas D (2000) Amyotrophic lateral sclerosis and invasive ventilation. In: Oliver D, Borasio GD Walsh D, eds. *Palliative Care in Amyotrophic Lateral Sclerosis*. Oxford, Oxford University Press, pp. 56–62.

Gelinas DF, O'Connor P & Miller RG (1998) Quality of life for ventilator-dependent ALS patients and their caregivers. *Journal of the Neurological Sciences,* **160** (Suppl. 1), S134–S136.

Holmes T. MND Association of England Wales and Northern Ireland. (2000) In: Oliver D, Borasio GD Walsh D, eds. *Palliative Care in Amyotrophic Lateral Sclerosis*. Oxford, Oxford University Press, pp. 143–7.

Kerkvliet J (2000) The role of the clinical psychologist in the ALS Team. In: Oliver D, Borasio GD Walsh D, eds. *Palliative Care in Amyotrophic Lateral Sclerosis*. Oxford, Oxford University Press, pp. 126–34.

Kingsnorth C (2000) Occupational therapy. In: Oliver D, Borasio GD Walsh D, eds. *Palliative Care in Amyotrophic Lateral Sclerosis*. Oxford, Oxford University Press, pp. 111–117.

Lyall R, Moxham J & Leigh N (2000) Dyspnoea. In: Oliver D, Borasio GD Walsh D, eds. *Palliative Care in Amyotrophic Lateral Sclerosis*. Oxford, Oxford University Press, pp. 43–56.

MND Advisory Group. (1999) *Guidelines for the Management of Motor Neurone Disease (MND)*. Endorsed by the Council of the. Association of Neurologists and circulated to members.

Mazzini L, Corra T, Zaccala M, Mora G, Del Piano M & Galante M (1995) Percutaneous endoscopic gastrostomy and enteral nutrition in amyotrophic lateral sclerosis. *Journal of Neurology*, **242**, 695–8.

Miller RG, Rosenberg JA, Gelinas DF, Mitsumoto H, Newman D & Sufit R (1999) Practice parameter: The care of the patient with amyotrophic lateral sclerosis (an evidence-based review). *Neurology*, **52**, 1311–23.

Mitchell JD (2000) Guidelines in motor neurone disease (MND)/amyotrophic lateral sclerosis – from diagnosis to patient care. *Journal of Neurology,* **247** (Suppl. 6), VI/7-VI/12.

Molloy I (2000) Rehabilitation. In: Oliver D, Borasio GD Walsh D, eds. *Palliative Care in Amyotrophic Lateral Sclerosis*. Oxford, Oxford University Press, pp. 135–42.

Neudert C, Oliver D, Wasner M & Borasio GD (2001) The course of the terminal phase in patients with amyotrophic lateral sclerosis. *Journal of Neurology*, **248**, 612–6.

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

O'Gorman B (2000) Physiotherapy. In: Oliver D, Borasio GD Walsh D, eds. *Palliative Care in Amyotrophic Lateral Sclerosis*. Oxford, Oxford University Press, pp. 105–11.

O'Gorman B & Oliver D (1998) Disorders of nerve I: motor neurone disease. In: Stokes M, ed. *Neurological Physiotherapy*. London, Mosby.

Oliver D (1996) The quality of care and symptom control – the effects on the terminal phase of MND/ALS. *Journal of the Neurological Sciences*, **139** (Suppl.), 134–6.

Oliver D (1998) Opioid medication in the palliative care of motor neurone disease. *Palliative Medicine*, **12**, 113–5

Oliver D (2000) Palliative care. In: Oliver D, Borasio GD, Walsh D, eds. *Palliative Care in Amyotrophic Lateral Sclerosis*. Oxford, Oxford University Press, pp. 23–8.

Oliver D & Webb S (2000) The involvement of specialist palliative care in the care of people with motor neurone disease. *Palliative Medicine*, **14**, 427–8.

Pegg R (1994) A carer's perspective. In: Williams AC, ed. *Motor Neuron Disease*. London, Chapman & Hall, pp. 203–12.

Scott A & Foulsom M (2000) Speech and language therapy. In: Oliver D, Borasio GD, Walsh D, eds. *Palliative Care in Amyotrophic Lateral Sclerosis*. Oxford, Oxford University Press, pp. 117–25.

Sykes N (2000) End of life care in ALS. In: Oliver D, Borasio GD, Walsh D, eds. *Palliative Care in Amyotrophic Lateral Sclerosis*. Oxford, Oxford University Press, pp. 159–68

Wagner-Sonntag E, Allison S, Oliver D *et al.* (2000) Dys-