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# Palliative care in motor neurone disease: where are we now?

David J. Oliver

**Abstract:** Palliative care has a very important role in the care of patients with motor neurone disease and their families. There is increasing emphasis on the multidisciplinary assessment and support of patients within guidelines, supported by research. This includes the telling of the diagnosis, the assessment and management of symptoms, consideration of interventions, such as gastrostomy and ventilatory support, and care at the end of life. The aim of palliative care is to enable patients, and their families, to maintain as good a quality of life as possible and helping to ensure a peaceful death.

**Keywords:** end of life, gastrostomy, motor neurone disease, non-invasive ventilation, palliative care

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## Introduction

Motor neurone disease (MND; also known as amyotrophic lateral sclerosis [ALS]) is a progressive neurological disease which leads to increasing paralysis of arms and legs together with speech and swallowing and breathing problems, due to progressive muscular weakness. The aetiology is unknown, although 5–10% of patients have a family history, and for these patients an abnormal gene mutation can be found in up to 60%.<sup>1</sup> In patients, with no family history, the same mutations are found in 10–20% and there is increasing evidence that there is a genetic component, with unknown environmental factors, that may lead to the disease.<sup>1</sup>

The prognosis for most patients is 2–3 years, although 25% of patients are alive at 5 years and 5% at 10 years.<sup>2</sup> The progression of the disease is very individual, but there is usually progressive loss of muscle power, which may affect the limbs, the bulbar area – affecting speech and swallowing – and the respiratory muscles of the chest wall and the diaphragm. The usual cause of death is respiratory failure, often associated with infection. There are now two drugs licensed for MND – riluzole, which has been shown to slow the progression in some patients,<sup>3</sup> and edaravone, which has been shown to help certain patient groups and is licensed in the United States.<sup>4</sup>

As there is, at present, no cure, palliative care may be seen as appropriate from the time of diagnosis, particularly as the prognosis is poor. Many patients may not be diagnosed for up to 12 months after their first symptoms and thus may have developed severe disability and may have a short prognosis.

There are many new developments and challenges in the care of a person with MND, and their family – in the communication about the disease, the use of interventions in the management of symptoms, the issues of exercise and physiotherapy, the psychosocial issues faced by patient and family and care at the end of life. The issues discussed below are of particular significance for patients and families and have been highlighted within national and international guidelines on the provision of care in MND.

## The role of palliative care

There has been increasing awareness of the role of palliative care for neurological patients<sup>5</sup> and in particular MND. Over the past few years, there have been developments in the care of patients with MND – in both the use of interventions, the provision of care, the use of guidelines and the care at the end of life, which may result in the need for careful assessment and discussion with

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patients and families, when a palliative care approach is helpful.

Many guidelines stress the need for palliative care including the European Federation of Neurological Societies,<sup>6</sup> the US Practice Parameter<sup>7</sup> and the National Institute of Health and Care Excellence (NICE) Guideline NG42 Motor neurone disease: assessment and management.<sup>8</sup> There is also increasing discussion of the need for close collaboration between neurology and palliative care services, with increasing education of all involved.<sup>5</sup>

In the past, palliative care was often only provided in the later stages of disease progression, near to death. However, palliative care is now increasingly seen as becoming involved related to the needs of the patient and family, rather than estimated prognosis.<sup>9</sup> Thus, a patient with MND may need extra support at varying times through the disease progression – at diagnosis when they face an unknown disease, as mobility is affected, when gastrostomy is discussed, when respiratory function is declining and non-invasive ventilation (NIV) is discussed and towards the end of life.<sup>10</sup> There may be periods between these occasions when the needs are reduced and the multidisciplinary team (MDT) can continue to support both patient and family. The use of triggers, which have been found to increase as death approaches, may help the end-of-life phase to be identified and may be helpful in helping all involved – patient, family and professionals – to be aware that death may occur in the coming weeks or months.<sup>11</sup>

There is increasing evidence for the effectiveness of palliative care in MND. A study, in a mixed group of 50 patients with neurological disease, including 16 people with MND, in Turin, Italy, showed that the involvement of the palliative care team improved quality of life and there was clinical improvement in symptoms – pain, dyspnoea, sleep disturbance and bowel symptoms.<sup>12</sup> Moreover, there was no increased mortality in the intervention group, compared with a control group, showing that palliative care was safe as well as effective.<sup>12</sup> Studies on MDT care, which will be described later, have also shown that a palliative care approach is helpful in improving quality of life and prognosis.<sup>13,14</sup>

There is also increasing evidence that palliative care is effective in other neurological diseases, which may support the involvement in MND. In the care of people with multiple sclerosis (MS),

studies have shown that there is an improvement in symptoms and caregiver burden with early short-term palliative care involvement<sup>15</sup> and this involvement was cost effective.<sup>16</sup> A further study which involved the education of specialist MS nurses to provide extra care did not show an effect on quality of life, but did show an improvement in symptom burden.<sup>17</sup>

Systematic reviews of palliative care for all diagnoses have shown small positive effects on quality of life and symptom burden, although many of the trials in the meta-analysis were assessed at having a high risk of bias.<sup>18,19</sup> Other studies have suggested that home-based palliative care for people with frailty, advanced heart failure, chronic obstructive pulmonary disease (COPD) on home oxygen, metastatic cancer and severe dementia may reduce admissions to hospital, with reduced healthcare costs.<sup>20</sup> A home-based service in Australia has shown that there are reduced hospital admissions and bed days and reduced costs of acute care in the last year of life.<sup>21</sup>

Thus, there is increasing evidence that palliative care should be considered for people with MND and the NICE Guideline recommended that palliative care expertise, either expertise of an existing MDT member or a specialist palliative care professional, should be an integral part of the MDT.<sup>8</sup> There are still barriers to this involvement as there may be fears that the palliative care team may have insufficient experience in MND care, the load on nursing care may be seen as excessive with insufficient resources to meet the need, or beds may become ‘blocked’ within specialist palliative care units.<sup>10,22</sup> Moreover, many patients, families and professionals associate palliative care with the end of life and the services may be viewed negatively, as they are associated with impending death.<sup>10,23</sup> There is challenge in explaining the earlier role of palliative care to all concerned.

### The telling of the diagnosis

As MND has a poor prognosis, palliative care may start from the telling of the diagnosis. This also provides the opportunity to set the agenda for later care. For instance, if the patient and family are not given accurate information or receive mis-information, such as ‘you will die choking’, many fears and anxieties may be engendered.

The NICE guideline recommends that the diagnosis should be given by a consultant neurologist

with experience and up-to-date knowledge of MND and its treatment and care.<sup>8</sup> The initial discussion about MND should be followed up by a member of the MDT, so that further questions can be answered. A clear single point of contact with the MDT was recommended to allow patients the opportunity to contact the team for further advice and support. The general practitioner of the patient should also be informed, so that everyone is aware of the situation.

These recommendations have been supported by recent research. In Australia, a survey of neurologists, patients and families looked at how the diagnosis was told, and compared the approaches to the SPIKES protocol, which has been suggested as a good way of breaking bad news.<sup>24,25</sup> In total, 36% of the patients who responded were dissatisfied with delivery of the diagnosis. The neurologists were graded as those showing high or low skills and the main areas where there were differences were responding empathetically to the feelings of patients and families, sharing information and suggesting realistic goals, exploring what patients and families were expecting, and making a clear plan and following this through.<sup>25</sup> In the Netherlands, a study showed that the organisation of two appointments, 10–14 days apart, helped patients and families to cope with the diagnosis.<sup>26</sup> The first appointment provided information, the intervening time provided the opportunity to re-orientate themselves to the new perspective on life and the second appointment allowed further discussion of the future plans and for the introduction of the MDT.<sup>26</sup>

Thus, a good approach to the telling of the diagnosis allows a partnership approach to be developed with the MDT so that both patient and family feel supported. This will help to establish the trust for future care and is a very important part of the palliative care of a person with MND. However, in a busy neurology clinic, the establishment of these norms may be challenging.

### **MDT care**

The role of the MDT in the care of patients with MND has become clearer and does seem to benefit care. There have been several studies that have shown improvement in symptom management and prognosis when an MDT is involved. In one study, patients under the care of a hospital clinic-based MDT were compared to historical controls, and the survival was found to be longer

for those with MDT care - 19 months – compared to 11 months for the control group.<sup>13</sup> Although the effect was augmented by the increased use of riluzole, NIV and gastrostomy, there seemed to be an independent effect of the MDT, although the exact nature of this is unclear.<sup>13</sup> In a further study in Ireland, patients were compared between the Republic of Ireland, where an MDT approach is present, and Northern Ireland, where a network approach occurs. There was a survival benefit of the MDT approach, with a survival advantage of 8 months, which again was not explained by the increased use of interventions.<sup>14</sup> The studies are both open to criticism as they were not randomised controlled trials and the control groups may not have been completely comparable. However, there does appear to be increasing evidence for the effectiveness of the MDT approach.

The review of evidence by the NICE guideline showed that there was a benefit of the MDT approach and an economic assessment showed that the incremental cost-effectiveness ratio was £26,672 per quality of life year (QALY) gained. This was considered to be a cost-effective intervention for a rare disease and was strongly recommended.<sup>8</sup> The MDT should involve several disciplines and include members with experience of MND care – in particular neurologist, specialist nurse, dietitian, physiotherapist, occupational therapist, respiratory professional, speech and language therapist and palliative care expertise, either from a trained member of the MDT or a specialist palliative care professional.<sup>8</sup> Thus, palliative care is seen as important from diagnosis and as an integral member of the MDT.

The importance of the MDT has been shown in these studies, but it is unclear how this is so effective. The MDT, as recommended by NICE, should be seeing patients regularly, every 2–3 months, and more often if there are problems and less often if the disease is stable. These assessments allow all areas to be considered and all members of the MDT to work in a co-ordinated way. This regular assessment and involvement of the wider MDT would seem to be important and further research is needed to elucidate the reasons for MDT effectiveness.

The need for continuity of care was also stressed within the NICE guideline. As the disease progresses, there will be the need for continual reassessment and adaptation to these changes.

Patients and families benefit from the MDT approach as the same professionals become involved as the disease progresses, rather than a new professional on each occasion.

### Cognitive change

There is increasing evidence of the presence of cognitive change in MND and that nerve loss is not only found in motor neurones but within the brain, and in particular the frontal lobes. Up to 50% of patients with MND may have cognitive impairment<sup>7,27</sup> and this may be seen as:

- Fronto-temporal dementia (FTD) – for up to 15% of patients,<sup>27</sup> and especially those with the C9orf 72 gene mutation (which is also associated with FTD alone). There is breakdown in cognition, behaviour and personality. Memory may be affected in varying ways but as FTD progresses there are increasing issues – with socially inappropriate behaviour, including disinhibition and gluttony – impairing everyday functioning.<sup>28</sup>
- ALS bi – behavioural impairment, seen in up to 28% of patients<sup>7</sup> with evidence of frontal lobe dysfunction. Changes in behaviour such as self-centeredness, irritability, apathy, emotional blunting, loss of embarrassment, disinhibition, lack of concern for themselves and reduced empathy may be seen.<sup>28</sup>
- ALS ci – cognitive impairment occurs in up to 39% of patients<sup>7</sup> and may be seen with behavioural changes. These changes may be found on testing, but there is increasing evidence that they are also clinically relevant. The changes are often in executive functioning – including organisation and planning, mental flexibility and reduced abstract thinking.<sup>29</sup>

Language impairment may also be seen in some patients, with difficulty in generating thoughts or finding words. There is increasing evidence for changes in emotional processing and the theory of mind – where patient may be less able to infer the mental state of other people or recognise emotions in others.<sup>27</sup>

All these changes have a profound impact on care. The recognition of these changes may be complex, and initially, they may be subtle and not appreciated. Families may be aware of changes in

behaviour and have concerns, and their involvement in recognition of cognitive change is important. There are several scales that can be used and the Edinburgh Cognitive and Behavioural Screen (ECAS) allows an assessment of the various aspects of MND cognitive change in 15–30 min and includes the questioning of families as well as the patient. The ECAS has been shown to be an effective screening tool and there are now alternative versions so that serial assessment may be possible.<sup>30</sup>

The recognition of cognitive change is important as this may affect the management and the consideration of treatment and interventions. It is thought that cognitive change may be seen early after diagnosis and so early screening can be helpful in future planning. Decision making may be affected if there is profound cognitive change, especially as executive function is affected, and so advance care planning may be helpful earlier in the disease progression, while the person has the mental capacity to make decisions. Whenever decisions are made, it is important to consider capacity and cognition, as some patients may find decision making very difficult and careful discussion when decisions are made will be needed. A best interest decision may be needed to be taken by the lead practitioner, in conjunction with the MDT, patient and family.<sup>31</sup>

There are increased stresses on the family and carers when coping with FTD or cognitive change. Caregivers report increased depression and anxiety and caregiver burden is increased. This is affected by behavioural change and physical impairment.<sup>32</sup> Explanation is needed to help caregivers, family and professionals understand the behaviour and to undertake strategies to cope with the changes, such as limiting decisions to two clear options.<sup>27,32</sup>

Professionals need to consider cognitive function when discussing management, particularly in the consideration of interventions such as NIV or gastrostomy. Cognitive change may make it very hard for a patient to tolerate any intervention and a Dutch study showed that patient with behavioural changes received NIV less often than other patient groups.<sup>33</sup> Moreover, the survival of patients with frontotemporal cognitive change was reduced compared to other patients – with a median survival of 3.4 years compared to 5.6 years in ALS without frontal changes.<sup>33</sup>



## Interventions in MND

Over the last 20 years, there has been increasing use of interventions in MND, in particular NIV and gastrostomy. These have a significant effect on quality of life and maybe length of life, but also raise ethical issues as the disease continues to progress.

### *Ventilatory support*

The majority of patients, as they deteriorate, develop weakness of the diaphragm and respiratory chest wall muscles. This may cause dyspnoea and will lead to respiratory failure.<sup>34</sup> The initial evidence for respiratory failure will often be at night, when the diaphragm is less able to move easily when lying down and the respiratory muscles may be more relaxed, causing the symptoms of orthopnoea, poor sleep, regular awakenings (due to episodes of low oxygen as a result of hypoventilation), excessive dreaming (as arousal interrupts rapid eye movement [REM] sleep), morning headache (due to carbon dioxide retention), feelings of never being refreshed, anorexia and even personality change.<sup>34</sup> On examination there may be signs of increased respiratory rate, orthopnoea, weak voice and cough, the use of accessory muscles of respiration, reduced chest wall movement and paradoxical movement of the abdomen.<sup>34</sup> More rarely patients may present with respiratory muscle weakness and even in respiratory failure.<sup>35</sup> Initially, the symptoms and signs of MND are often missed, often due to misdiagnosis.<sup>36</sup> The diagnosis of MND may only be made after initial resuscitation, and ventilator support has been commenced.<sup>35</sup> This occurs rarely and usually patients remain ventilator dependent.<sup>35</sup>

NIV has been shown to be effective in reducing symptoms, improving quality of life and extending life. There has been one randomised controlled trial showing an improvement in several measures of quality of life and a median survival benefit of 205 days – those receiving NIV had a median survival of 216 days opposed to 11 days who just received best supportive care.<sup>37</sup> However, not all patients are able to tolerate NIV and may experience problems with the use of technology, sleep disturbance, the sensation of pressure and pulsing, dry mouth and mask concerns, including claustrophobia while wearing the mask.<sup>38</sup> However, many patients and families do find NIV to be beneficial, with increased energy, improved sleep, improved breathing, better speech and improvement in caregivers' well-being.<sup>34,37</sup> Patients with bulbar issues were found to have a poorer outcome when using

NIV<sup>37</sup> and those with cognitive change or FTD may have considerable difficulty in coping with the equipment and the restriction with NIV.<sup>39</sup>

The discussion and consideration of NIV is complex, and palliative care professionals may be able to offer the opportunities for these discussions. Moreover, there is the need to discuss what will happen in the future, for although these symptoms of respiratory failure may improve, the disease will continue to progress, with increasing disability. It is very helpful for these discussions to take place early in the disease progression, before there is an urgent need to start NIV, so that time can be taken in the discussion and decision making. The NICE guideline suggests that the use of NIV should be discussed when respiratory function is assessed, and this should start from early in the disease and soon after diagnosis so that early changes can be detected.<sup>8</sup> The Guideline also recommends that when NIV is started, there is the need to discuss the advantages and disadvantages of NIV and the possibility of becoming dependent on NIV, when the disease has progressed with increased disability.<sup>8</sup> Discussion of the future and the future planning is essential so that patients can state they would wish if there was further deterioration,<sup>8</sup> as advance care planning – which ensures the patient's wishes are known, while they have capacity, so that if capacity is lost later their wishes can be respected.<sup>31</sup>

As the respiratory function continues to deteriorate, the use of NIV may increase. Initially, the use is at night, when the patient is lying down, but there may be a need for NIV during the day and over time some patients become totally dependent on NIV and are only able to tolerate removal for a short time, for eating or hygiene care. This may lead to complex decision making, as the patient may ask to stop NIV. If they are dependent on NIV, removal could cause severe symptoms of breathlessness and distress<sup>40</sup> and so medication is often needed to relieve distress and manage breathlessness before withdrawal.<sup>40</sup> Patients who are not 24 h dependent may still experience distress at withdrawal and require medication.

Palliative care services may have a role in helping to facilitate the discussion and decision making at this time. If the patient has full capacity to make decisions, then there are no ethical or legal barriers to stopping this intervention, in the United Kingdom and many other countries. If a patient does not have capacity for the decision, as may

occur with cognitive change of FTD, a 'best interests' decision may need to be made by the lead medical practitioner, in consultation with the wider MDT and the family. The MDT may also need to take into account any advance care plan made by a patient earlier, when they did have capacity, and in particular a well-defined Advance Decision to Refuse Treatment (ADRT) which may clearly define when a patient would wish to stop NIV.

However, there are many practical aspects to be considered, and although the action is ethical for many people, the cessation of a treatment, and the ensuing death of the patient within a short time, can feel as if they have caused the death. A survey of palliative medicine consultants in the United Kingdom found that they found the discussion of the procedure, the varying team reactions and the practical, ethical and emotional aspects all very difficult. They were asked to rate how challenging they found these decisions, with a scale from 1 - not at all challenging - to 10 - very challenging. In total, 32% rated the practical aspects at over 7 out of 10, 33% rated the ethical aspects at over 7 out of 10 and 49% rated the emotional challenge at over 7 out of 10.<sup>41</sup> Further studies have confirmed that this is a difficult and complex area for professionals<sup>42</sup> and the Association for Palliative Medicine of Great Britain and Ireland has produced a guidance for professionals, in association with other professional bodies.<sup>40</sup> This outlines the ethical and legal position and provides a clear plan for the procedure and support from mentors who have been involved in the withdrawal of NIV in the past.<sup>40</sup>

One challenging and developing area is the increasing discussion of the role of tracheostomy and invasive ventilation. In the United Kingdom, this is rare and often only occurs before the diagnosis is known, such as when a person presents in acute respiratory failure. Some patients may wish to consider tracheostomy as an elective procedure, particularly if they have severe problems with secretions or they cannot tolerate NIV. There is evidence that tracheostomy tends to be chosen by younger patients and those who are wanting to stay alive.<sup>43</sup>

The rate of tracheostomy across the World varies greatly, with the United Kingdom at only 1–2%, in some areas of Italy 10–30%<sup>44</sup> and in Japan over 30% of all patients with MND have a tracheostomy and many live for up to 10–20 years.<sup>34</sup> Although tracheostomy may reduce the problems

of secretions and aspiration, there is the risk of increased disability, reduced communication and even, in up to 15%, becoming 'locked in' with no way of communicating.<sup>45</sup> Moreover, there is evidence that although the quality of life of the patient may be maintained, many caregivers find the burden very great, and in one study 30% of carers rated their own quality of life lower than that of the patient.<sup>46</sup> The prognosis after tracheostomy is unclear, but in European studies, the median survival was 24 months in Italy<sup>44</sup> and 56 months in Denmark<sup>43</sup> but with a large range in all areas. The issues of withdrawal of ventilation may have to be faced, as with NIV, and the death will usually occur shortly after ventilation is removed.

There are many issues that need to be considered, and it is very helpful for these discussions to start before a decision is needed, so that time can be taken for discussion and consideration of the positive and negative aspects. The NICE guideline suggests that monitoring of respiratory function should be undertaken from diagnosis and that discussion of the role of NIV, the benefits and drawbacks can be introduced over months and years, rather than a rushed decision when someone is very breathless, distressed and intervention needs to take place urgently.<sup>8</sup> All too often tracheostomy may occur in these urgent scenarios and be regretted later.<sup>47</sup>

#### *Cough management*

Respiratory muscle weakness affects the ability to have an effective cough. Normally, the inspiratory muscles increase lung volume, expiratory muscles produce the increased thoraco-abdominal pressure and the upper airway muscles co-ordinate glottis closure and opening, allowing a cough. All of these aspects may be affected to a varying degree in MND.<sup>48</sup> A reduced cough reduces the clearance of secretions and increases the risk of respiratory infection. It has been suggested that a peak cough flow of 160 L/min is necessary to clear the airway, and if patients' peak cough flow is less than 270 L/min, they should be taught cough augmentation techniques, to ensure that they can clear debris and cope with an infective episode, when the cough is usually reduced.<sup>48</sup>

There are several techniques that can be used and the NICE guideline recommends that manual assisted cough techniques should be offered to patients who cannot cough effectively.<sup>8</sup> Breath stacking or assisted breath stacking, using a lung

volume recruitment bag, is recommended as the initial treatment options, although there is little evidence in the literature.<sup>8</sup> These techniques allow the patient to maximise the volume of air they can inhale and then to cough with an increased cough pressure.<sup>49</sup> There may be difficulty if there are bulbar issues and the patient and family will need careful education in the technique.

The use of a mechanical insufflator-exsufflator has also been suggested.<sup>34</sup> There is little evidence that this is more effective for many patients and it is expensive and some patients find the pressures difficult to cope with. The NICE guideline does recommend considering a mechanical cough assist device if breath stacking is not effective or during a respiratory tract infection.<sup>8</sup> This has been supported by a study showing that there is deterioration in respiratory muscle strength in an infective episode and the mechanical insufflator-exsufflator was effective for 83% of patients in an infective episode, compared to 35% with manually assisted coughing.<sup>50</sup>

Careful assessment of the ability to cough, and the peak cough flow is important, particularly for the patient starting on NIV. The use of techniques to aid coughing and the consideration of a mechanical insufflator-exsufflator may be necessary, particularly during an infective episode, and may prevent hospitalisation.

### *Gastrostomy*

The maintenance of nutrition in the care of a person with MND has developed over recent years. The assessment of the nutritional status of patients is important and should be part of the regular MDT assessment, including measurement of weight and the assessment of diet and swallowing, nutritional input, fluid intake, oral health and swallowing.<sup>8</sup> There is a real challenge to professionals to be more aware of the nutritional state of patients, as even weight loss of 5% appears to be associated with increased mortality.<sup>51</sup> Further studies are being undertaken to look at the benefits or risks of increased feeding, even in the early stages of the disease. The concerns about nutrition have increased the consideration of interventions to maintain weight and well-being.<sup>52</sup>

Reduced nutritional input may be due to swallowing issues but also could be related to arm weakness (if that the person is less able to feed themselves), respiratory issues (with breathlessness

on eating), or depression or low mood. An MDT assessment will allow for the provision of eating aids to help the person take food and drink into the mouth or for help with the preparation of food, so that it remains nutritious and is also appealing and tasty. Advice may also be necessary to help with the positioning, seating and posture for meals, as neck weakness may cause swallowing to be difficult. Alternative feeding may be considered, often using food in a 'custard' consistency or the use of thickeners or nutritional supplements. The role of eating in social situations needs to be considered, as issues of swallowing or in eating may lead to the person eating less due to embarrassment in social situations, such as at a family meal or when eating out. Fear of aspiration or choking may affect a person eating, and a wider discussion may be required.<sup>8</sup>

The use of gastrostomy – as a percutaneous endoscopy gastrostomy (PEG), percutaneous radiologically inserted gastrostomy (PRG) or per-oral image-guided gastrostomy (PIG) – has increased over the last few years. There is mixed evidence of the effectiveness of gastrostomy on survival, but there is evidence that there may be a positive effect on quality of life and survival.<sup>52</sup> The timing of gastrostomy insertion is complex. The risks of insertion are related more to respiratory function than swallowing or nutritional aspects, as there are increased risks of insertion of a PEG if the forced vital capacity (FVC) is less than 50% of expected.<sup>7</sup> The concerns may be less if a PRG or a PIG is used, as sedation may not be necessary and the patient does not need to be recumbent.

These discussions about gastrostomy may need to start when swallowing is starting to cause problems and respiratory function is deteriorating.<sup>8</sup> This may feel too early for the patient but an earlier insertion is safer and less likely to lead to complications. The gastrostomy does not need to be used initially but may be used over time to supplement, rather than totally replace, oral feeding, particularly when swallowing is more difficult, such as during an infection. The use may increase slowly and be used as required. A gastrostomy may also facilitate care at home, as nutrition, hydration and medication can be continued more easily, which may allow a patient to remain, and die, at home.<sup>53</sup>

The discussion may be complex. A study has shown that people who were more likely to make decisions compared to others were found to have a higher IQ, longer education, an 'active approach'



to gastrostomy and NIV and actively seeking information, services and interventions.<sup>39</sup> Another important factor in decision making is the patient's pleasure in eating and it is very important to stress to patients that some oral feeding, for taste and pleasure, can continue with the gastrostomy used for supplementary feeding. Patients with evidence of cognitive dysfunction were also less likely to accept a gastrostomy and so careful assessment of cognition should be included when decisions are made.<sup>39</sup> The study also shows the importance of ensuring that patients and families do understand the decisions and the implications of an intervention, with extra care needed if the education level, IQ, understanding of the disease and social support are reduced.<sup>39</sup> Medical and allied healthcare professionals may hold different attitudes to interventions and this can affect decision making. Thus, there is a challenge to ensure nutritional aspects are assessed and options discussed appropriately. These discussions do need to be undertaken earlier in the disease progression, so that there is time for the decision process and a sudden decision, during a crisis or when swallowing has become very difficult, is avoided.

#### **Weakness and the role of exercise**

A person with MND faces increasing muscle weakness and reducing functional ability and challenges the professional team. There is little evidence that physiotherapy or other activity can prevent these changes, although resistance exercise and range of motion exercise may reduce pain, cramps with some improvement in function, as measured by the ALSFRS scale.<sup>54</sup> Resistance and endurance exercise have also been shown to be safe and does not worsen function.<sup>55</sup> The NICE guideline has recommended exercise programmes to maintain the range of joint movement, prevent contractures, reduce stiffness and discomfort and to optimise function.<sup>8</sup> Family members and carers should help in any exercise programme and the patient and family may also benefit from advice on safe manual handling.

Neck weakness may cause particular distress to patients, as this may cause pain across the neck, affect breathing, communication and eating, increase drooling and affect quality of life, as the patient is less able to look at their environment and be involved in social contact. There have been many different collars to help with this head drop but all have problems – reducing mobility, increasing stiffness and pain and they are often

very obtrusive. A new collar – Head-up – has recently been developed following a careful assessment and development project.<sup>56,57</sup> This does support the neck but can be adjusted to cope with the individual's needs, is less likely to cause discomfort and is aesthetically more acceptable.<sup>56,57</sup>

#### **Psycho-social aspects of care**

The psychological support of the person with MND is very important. They face multiple, continual loss – of speech, swallowing, movement, mobility, cognition, emotional expression, social interaction – and face dying and death. The MDT should discuss the emotional and psychological aspects of care at all appointments,<sup>8</sup> and this may include the wider understanding of MND and its effects on daily living, the acceptance of the diagnosis, adjustments to changes in work, lifestyle and their usual activities, changes in family roles and sexuality/intimacy and their own concerns about their family. Ongoing support is essential, and the opportunity to discuss these issues is important, with someone who does know about MND and its implications.

There would seem to be no specific psychosocial interventions for people with MND and a study of a group of therapists suggested that their main aims were to support the person in the 'here and now', focus on what they were able to do, reaffirm the person in their ability to maintain a role in life and support the person in exploring their emotions.<sup>58</sup> It is important for the therapist to have an understanding of MND and how it may affect someone.<sup>58</sup> The provision of clear information, clear communication and addressing fears and uncertainty are important.<sup>59</sup> A MND Advisory Service in Australia provided home visits and telephone support for patients and families and was greatly valued, with both groups feeling more supported and more able to make informed decisions.<sup>60</sup>

Depression is seen more commonly in MND, particularly in patients over 65 years and in the first year after diagnosis.<sup>61</sup> Rates in studies vary from 4.4%<sup>61</sup> to 12%.<sup>62</sup> The use of antidepressants was also higher in MND, although tricyclic antidepressants may also be given to help with drooling. Although the wish to die was common, with 25% of patients expressing these wishes, the majority did not have depression, with only 37% of this group showing evidence of clinical depression.<sup>62</sup>

Family caregivers have been shown to have many issues, which are often not recognised. They may have to cope with many areas, particularly as the disease progresses. A study of caregivers in Norway showed that they felt there were five predominant areas of concern – immediate care work, seeking information about MND, managing competing obligations, maintaining normality, and managing external resources and assistance.<sup>63</sup> It was complex to balance all these areas and issues, while providing increasing care for the person with MND.

There is appreciable burden of care on family caregivers. This may be increased if there are social restrictions due to caring for the patient, self-criticism of their caring and anger and frustration at the various situations, often associated with guilt about the situation they face in caregiving.<sup>64,65</sup> Moreover, there is increasing evidence that these burdens are greater when the patient is more disabled physically and there are behavioural issues, associated with cognitive change.<sup>32</sup>

The awareness of possible caregiver burden is important in providing care to both patient and family. It is important to ensure that interventions do not increase the burden of carers, this again emphasises the importance of ensuring that there is consistent care from caring support services.<sup>8</sup> The NICE guideline recommended that social care should be undertaken by people who know about the disease and this care should continue with a small team of people known to the patient and family, to allow continuity of care and familiarity with the needs of the patient.<sup>8</sup> Many families feel that they spend most of the time they receive from carers in explaining the issues and needs of the person. This is reduced by the provision of a small, consistent team.<sup>8</sup>

It may be useful to undertake regular assessment to identify family support needs and thus allow the most appropriate and helpful support. The Carer Support Needs Assessment Tool (CSNAT) has been shown to identify the priority areas for support and the tool was well accepted by carers.<sup>66</sup> A study using CSNAT showed that the main priorities were support for future care, having a contact when concerned and the provision of equipment to help in care.<sup>10</sup>

Thus, the wider assessment of the whole patient and whole family, which is an implicit part of palliative care, is a vital part of the care of someone

with MND and continual assessment and discussion is needed.

### End-of-life care

As MND has no curative treatment and has a prognosis for many of only 2–3 years, end-of-life issues may need to be considered early in the disease progression, particularly as there may be delays in diagnosis so that the disease has progressed by the time the diagnosis has been made. There is evidence that patient and families do appreciate the discussion of end-of-life issues<sup>10</sup> and the NICE guideline recommended that end of life should be discussed if the patient or family ask and when considering and commencing new interventions, such as gastrostomy and NIV.<sup>8</sup>

However, there is often reluctance to discuss the issues of dying and death, although many patients and families have great concerns about the future. There are many myths that dying of MND is ‘distressing’ and ‘horrendous’ and is ‘due to choking’. There has been widespread publicity when certain cases are discussed in the UK Courts, when people have asked for the right to an assisted death.<sup>67</sup> There is increasing evidence that death for MND is usually peaceful and studies have shown that choking of death is rare – in a study of patients at St Christopher’s Hospice in London only one patient died choking, and this was in 1968 soon after the hospice opened;<sup>68</sup> in a further study of 121 patients in the United Kingdom and Germany, no patient died choking and over 88% patients died peacefully.<sup>69</sup>

These myths need to be confronted and evidence provided that death can be peaceful.<sup>68,69</sup> A study in France has shown that the majority of patients died of respiratory failure (77%) and 10% died of other causes – post surgical or trauma (5%), cardiac disease (3.4%), suicide (1.3%) or unknown causation (0.7%).<sup>70</sup> However, there were many patients, in particular those dying outside of hospital, where the cause could not be elucidated. The use of interventions had continued until near to death in many patients – 55% were still taking riluzole, 33% were on NIV and 37% had a gastrostomy in use.<sup>70</sup> In Hong Kong, a study of 52 patients recorded the main causes of death as pneumonia (54.8%) and respiratory failure (40.5%).<sup>71</sup> Thus, the evidence does show that death is usually peaceful and a distressing death is rare – and no different for dying from other diseases.

However, the issues of dying and death should be discussed with patients, particularly as interventions such as NIV and gastrostomy are introduced.<sup>8</sup> As discussed above, the withdrawal of treatment may need to be considered – at the patient's request or in response to advance care planning. Patients and families may wish to discuss these issues and both patients and families may vary between acceptance and hope.<sup>60</sup> The opportunity for families to talk and share experiences together is important and facilitating this sharing may become the role of anyone in the MDT.<sup>72</sup>

Caregivers may have specific issues. It has been suggested that the issues that family caregivers face are the conflicts of trying to prepare for the future while they feel overwhelmed by the issues of caring throughout the illness progression and coping with uncertainty.<sup>10</sup> Family caregivers talked of the themes of coping with the 'Here and Now' – with the privilege of caregiving, the all-consuming nature of care, coping with family cohesion and conflict and the difficulties of coping with variable health and social care support; negotiating with the Here/After – with a unpredictable trajectory of disease progression, trying to prepare but being unable to plan and preparing while living with both hope and fear and feelings of self-reproach when thinking of the future.<sup>10</sup> Thus, many caregivers may be prepared cognitively, some were prepared behaviourally but many found preparing emotionally challenging. Thus, professionals should not assume that all are well prepared for the future, and death, and need to carefully assess each family.<sup>10</sup>

The discussion of future care, including dying and death, will also enable the opportunity for advance care planning – expression of the person's wishes for future care which can influence decisions if the person loses the ability to make the decision themselves, due to loss of communication or cognition. This is particularly important to consider for people with MND as both communication and cognition may be affected, particularly near to the end of life. The person may express their wishes as to the interventions they may not wish, such as cardiopulmonary resuscitation, tracheostomy, gastrostomy, ventilator support, place of care or death, their financial wishes in the form of a will, funeral arrangements or the overall care at the end of life, such as specific wishes for music or family/friends to be present at the time of death.<sup>31</sup> In the United Kingdom, a person can make a specific ADRT, which if

specific and stating that the person realises that the decision could lead to their death is legally binding, or a Lasting Power of Attorney, where other people are chosen, who would make decisions on the person's behalf.<sup>31</sup> These only apply if the person has lost capacity to make the decision, for if they are able to decide for themselves they should be enabled to make the decision. Other aspects of care at the end of life may also need to be considered, and it is important to ensure that there is clear understanding about these issues, as for instance there is evidence that place of care and place of death may be seen as separate decisions and need to be clarified clearly and separately.<sup>73</sup>

Some patients with MND may wish to discuss hastening of death – by euthanasia or physician-assisted suicide. In countries where this is legal, there is evidence that people with ALS do chose an assisted death more often than other people – in Oregon the rate of physician-assisted suicide was 374.5/10,000 compared to 61.0/10,000 for cancer<sup>74</sup> and in the Netherlands about 20% of patients with ALS receive euthanasia compared to 5% of cancer patients or 0.5% of heart failure.<sup>75</sup> When a patient does discuss hastening death, this is an opportunity to discuss the reasons behind the request – which may be fear of being 'kept alive' by ventilation, fear of the dying process or fear and concerns about death itself.<sup>23</sup> It may be possible to address some of these issues and the desire for a hastened death may reduce.<sup>23</sup> However, some people may wish to strongly express their autonomy, and in legislations where no action can be taken, there is a continued need to support the patient and family and see if any further discussion may be helpful.

There is often the need to recognise when the end of life is approaching for someone with MND. The deterioration may be rapid, often with an infection or a sudden deterioration in respiratory function.<sup>11</sup> There is evidence that there may be triggers which may indicate when the end of life is near for neurological patients: swallowing problems, recurring infection, particularly aspiration pneumonia, marked decline in functional status, cognitive difficulties, weight loss and significant complex symptoms.<sup>11</sup> For people with MND, further triggers were suggested: respiratory failure or increased breathlessness, reduced mobility and dysphagia.<sup>11</sup> Although these triggers were agreed by expert consensus, studies have shown that the number of triggers increases nearer to death and

patterns of symptoms may be seen.<sup>76</sup> Thus, these triggers may be useful, in conjunction with a wider assessment of the patient, by identifying when death may be near, so that the care provided may be adjusted – such as careful consideration of interventions such as continuation of NIV or gastrostomy feeding, provision of anticipatory medication, for use at home in a crisis situation, and fuller discussion with the patient, if appropriate, family and the wider MDT.

### Conclusion

All patients with MND will benefit from palliative care and all involved in patient care should be able to provide a palliative care approach – listening to the patient and family and assessing and managing issues – physical, psychosocial and spiritual.<sup>77,78</sup> Some patients and families may have more complex needs and a specialist palliative care team approach – from a specialist unit/hospice – may be helpful. This team will have greater expertise, have had specialist training and continuing education.<sup>77,78</sup>

There will need to be a collaborative approach involving many services – including primary/family/home care but many specialist services, such as gastroenterology, respiratory medicine, neuropsychology as well as neurology and palliative care services. It is important to ensure that these services provide co-ordinated care and many suggest a specific/key person or team with whom the patient and family can have regular contact.<sup>8,9</sup> It is also important to acknowledge that different teams will act in different ways and may have varying ways of collaboration and differing ethos;<sup>79</sup> if these differences are acknowledged, the care for the patient and family is likely to be better co-ordinated and effective.<sup>79</sup>

Increasingly, particularly in the United States, neuro-palliative care is developing aiming to provide the specialist MDT approach.<sup>80</sup> This may develop further, but in many areas, there will be the need to develop a close collaboration between neurology and palliative care so that all patients with MND can receive the most appropriate and effective care, maintaining quality of life and allowing a peaceful death.

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