Review issue

David Oliver
FRCP, FRCGP, Tizard Centre, University of Kent, Canterbury, Kent, UK

The development and implications of guidelines for care of people with amyotrophic lateral sclerosis

Abstract

The care of a patient with amyotrophic lateral sclerosis is complex and involved a wide multidisciplinary team. Recent guidelines in the UK have been produced by the National Institute of Health and Care Excellence and this paper looks at the recommendations and implications for practice.

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Key words: amyotrophic lateral sclerosis, care guideline, multidisciplinary care, symptom management, respiratory management

Introduction

The care of a person with amyotrophic lateral sclerosis (ALS, also known as motor neurone disease, MND) is complex. The disease is characterised by progressive neurodegeneration – primarily of motor neurones but affecting the frontal lobes and other areas of the brain and nervous system [1]. There is no known cause, although 5–10% have a family history and for the majority of these people an abnormal gene can be found [2]. There is increasing understanding that there is a genetic component with other factors causing the development of the disease [2]. The prognosis is usually of two to three years from diagnosis, although 25% are alive at 5 years and 5% at 10 years [1].

The person with ALS faces progressive disability — with weakness of arms and legs, speech and swallowing problems and respiratory muscle weakness, causing breathlessness and respiratory failure, which is the usual cause of death. The family and carers of the person with ALS also face multiple losses [3]. The aim of care should be to maximise the abilities the person has and to maintain the quality of life. It can be argued that palliative care is appropriate from the time of diagnosis and certainly the principles of palliative care – holistic assessment by a multidisciplinary team applied from diagnosis [4].

Within the UK there are different models of care for people with ALS. There are specialised MND Centres, often supported by the MND Association, but many people with ALS do not see specialised services and are not well supported through the disease progression. As a result, the National Institute for Health and Care Excellence (NICE) planned a Guideline on the management of people with MND. This involved the

Corresponding address: David Oliver
Tizard Centre, Cornwallis North East, University of Kent, Canterbury
Kent CT2 7NF, UK
tel: 01227 827373
e-mail: drdjoliver@gmail.com

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formation of a Guideline Development Group, supported by the technical staff of the National Guideline Centre of the Royal College of Physicians. The Group considered different aspects of care, looking at the evidence of the literature, using the GRADE system, and making recommendations for care [5]. If there was insufficient evidence, the recommendations were agreed by consensus discussion.

There have been previous guidelines for the care of people with ALS. In the USA there is a Practice Parameter, produced by the American Association of Neurology [6] and in Europe the EFNS Guideline on ALS [7]. In the UK there had been NICE Guidelines on the use of riluzole, the only medication licensed for the treatment of ALS, with a small survival advantage [8] and on the use of non-invasive ventilation [9]. The latter guideline was to be incorporated within this new NICE Guideline [10].

This paper focus on the main recommendations, and the implications for the care of people with ALS. Although this process took place in the UK the evidence was taken widely and the recommendations are relevant to other countries, subject to differences in health and social care systems.

Recognition and referral

There is awareness that ALS is a rare disease within the community and the diagnosis may be delayed. It has been recommended that there are clear protocols and pathway for referral. Awareness of possible symptoms is encouraged and the Royal College of General Practitioners and the MND Association have produced a document on "Red flags" with suspicion being raised if there is progressive muscle weakness [11]. It is also stressed that information should be provided for the patient and family at all stages.

Diagnosis

The diagnosis should be given by a Consultant Neurologist with knowledge and expertise in ALS. In emergency/urgent situations, such as diagnosis of ALS being made at the presentation with acute respiratory failure, other specialties may need to tell the diagnosis but further discussion should be with an experienced neurologist. It is recommended that patients are asked about their wishes for information and the involvement of their family and information should be provided as they wish. A single point of contact should be provided, with the ALS Multidisciplinary Team (MDT) and one of the Team should see them within 4 weeks of the initial appointment to discuss the issues that may arise for the patients and family.

Organisation of care

The main recommendation for the organisation of care is to be for a clinic based MDT. This would usually be based in a hospital setting, but could be in the community, and would include health and social care professionals with expertise in ALS care and working closely together. The membership would be:

- Neurologist
- Specialist MND nurse
- Dietitian
- Physiotherapist
- Occupational therapist
- Respiratory healthcare professional
- Speech and language therapist
- Palliative care expertise — this may be one of the team members. Referral to specialist palliative care should be considered if there are complex needs.

Patients would be seen at this Clinic every two to three months, but more often if there are increasing symptoms or issues and less often if there is slower deterioration. The Team would assess, manage and review the main areas of care:

- Weight, nutritional intake, feeding, swallowing
- Muscle problems — weakness, stiffness, cramps
- Physical function
- Saliva problems — drooling, thick saliva
- Speech and communication
- Cough effectiveness
- Respiratory function

These recommendations are based on several studies that have shown that not only the quality of life, but length of life, are improved with MDT care — one study showed one study suggesting the 1 year mortality was decreased by 29.7% [12], another suggesting that the median survival was 19 months for the MDT group compared to 11 months for standard care [13] and a further study showing a survival difference [14].

Other areas of care are considered:

- Psychological support
  The MDT assessment should include discussion of the psychological and emotional impact of ALS, and information on possible support systems should be provided.
- Provision of equipment
  The assessment by physiotherapy and occupational therapy should be reviewed regularly and include consideration of activities of daily living, the home environment and the need for technology. Any equipment should be provided without delay and should be able to be adapted, to cope with the person's deterioration.

- Assessment of cognitive changes
  As cognitive change in ALS is more widely understood — with 10–15% of patients having fronto-temporal dementia and a further 35% showing evidence of frontal lobe changes [15] — there is a need for discussion about cognition and behaviour with patients and their families from diagnosis or if there are particular concerns. A formal assessment may be necessary. If there is evidence of cognitive change, discussions about care may need to be carefully tailored to the patient's needs, considering their communication abilities, cognition and mental capacity to make decisions.
- Communication
  The needs for face to face, telephone and social media communication of a patient should be assessed and the equipment should both low-tech, such as a picture board or alphabet board, and high-tech, such as computer based technologies.
- Nutrition
  There is a need to assess weight, nutrition and swallowing from diagnosis. There may be issues with the ability to eat and drink – food may need special preparation, often a custard consistency is easier to swallow, or help may be need in preparation, aids may be needed to help the person take food from the plate to the mouth and advice may be needed on posture, positioning and seating for mealtimes. There may be the need for advice in coping with social situations — as eating is often a social and family activity. If swallowing problems occur, a careful swallowing assessment is necessary and discussion of the placement of a gastrostomy should start early and occur regularly. If a gastrostomy is required, this should be placed without delay.

Symptom management

A limited number of symptoms were considered:

- Muscle problems
  - Cramps

Cramps may occur in spastic muscles and the treatment is recommended;

  - First line — Quinine
  - Second line — Baclofen, tizanidine, dantrolene, gabapentin
  - It is important to ensure medication is appropriate, such as can be swallowed and taken and the use should be monitored carefully for effectiveness and side-effects
  - Physiotherapy

This may help maintain joint movement, prevent contractures and reduce stiffness and discomfort. Physiotherapy may be offered as resistance, active assisted or passive movements. Families and carers should be encouraged to assist in any exercise programme.

- Saliva
  Careful advice on posture, diet, swallowing and oral care is essential. Treatment may be
    A trial of a antimuscarinic — e.g. glycopyrrolate
    Injection of Botulinum toxin into salivary glands
    If saliva is thick, reduce or stop medication that may thicken saliva, advice on diet and posture and consider humidification, nebulisers or carbocisteine.
- Cough augmentation
  Cough augmentation techniques should be offered if the patient is not able to cough effectively:
    Breath stacking or manual assisted cough may be helpful
    If there are bulbar changes or breath stacking is ineffective, consider:
      Assisted breath stacking — using a lung volume recruitment bag
      Mechanical cough assist machine — particularly if breath stacking is ineffective or during respiratory infection

Respiratory function

There is the need for regular assessment of respiratory function, as the aim is to find early changes, before respiratory muscle dysfunction causes respiratory failure. This needs to be carefully explained to patients and families and if there is evidence of respiratory muscle weakness, causing symptoms such as orthopnoea, breathlessness, disturbed sleep or morning headache, non-invasive ventilation may need to be considered.

NIV may be discussed soon after diagnosis and when respiratory monitoring starts. Patients and families should be given appropriate information on the consideration of the benefits of the improvement of symptoms but also the risks of continued deterioration and dependency on NIV. Although there is strong evidence that NIV will prolong life [16] the underlying progression of ALS will not be altered.

NIV should be offered if there are symptoms and evidence of respiratory muscle weakness and may be started initially by acclimatisation in the day and slowly increasing the use at night. There is a need to undertake a risk assessment of how the patient and their family will cope with the NIV interface, and the risks of ventilator or power failure. This will include discussion about their wishes if there are further problems, such as infection or increasing dependency or symptoms. Medication to cope with breathlessness

and distress — morphine sulphate, midazolam, and an antimuscarinic, such as glycopyrronium bromide or hyoscine hydrobromide — may be provided, so that they are readily available if there is a sudden deterioration.

If a patient has frontotemporal dementia or severe bulbar problems, with severe drooling of saliva, NIV may not be easy to cope with. The use of NIV may need to be carefully assessed and may not be appropriate. If there are severe symptoms a trial of NIV could be considered.

Some patients on NIV will become dependent and even approach using the ventilator support for 24 hours a day. They may ask to stop the NIV and if the patient is able to make this decision autonomously it is ethical for this treatment to be withdrawn. However, a plan to withdraw is essential, as medication may be needed to ensure that they do not become distressed. Extra support is also needed for family and all the professionals in the team [17].

End of life care

There are clear recommendations that all professionals should be prepared to discuss end of life care whenever the patient and family ask and to provide advice on advance care planning, so that the patient's wishes are known if they lose capacity to make decisions later by themselves. These wishes may include their wish of place of care, place of death and what they would like to happen if they deteriorate suddenly or develop a severe infection or another illness or complication. Specialist palliative care services may become involved if they are not already seeing the patient and family.

As the end of life approaches extra help should be provided to allow the family and carers to spend more time with the patient and have some of the responsibility for care to be lessened. Extra equipment and care should be provided if they are not already present, such as communication aids, specialised beds, commode or a hoist. Bereavement care should be available for families after death.

Conclusion

These guidelines allow health and social care professionals to develop improved care for all people with ALS, using an evidence based approach. The challenge is now to ensure that the recommendations are implemented and patients and their families can experience the improvement in the care they receive.

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