Available online at
ScienceDirect
www.sciencedirect.com

Elsevier Masson France
EM|consulte
www.em-consulte.com

Motor neuron diseases

Care management in amyotrophic lateral sclerosis

M.-H. Soriani *, *C. Desnuelle*

Centre de référence maladies neuromusculaire/SLA, university hospital of Nice, CS 51069, 06001 Nice cedex 1, France

INFO ARTICLE

Article history:
Received 30 September 2016
Accepted 27 March 2017
Available online 29 April 2017

Keywords:
Amyotrophic lateral sclerosis
ALS management
Non-invasive ventilation
Nutrition
Palliative care
Ethics
Rehabilitation

ABSTRACT

Amyotrophic lateral sclerosis (ALS) is a relentlessly progressive and fatal neurodegenerative disease characterized by progressive weakness of voluntary muscles of movement as well as those for swallowing, speech and respiration. In the absence of curative treatment, care can improve quality of life, prolong survival, and support ALS patients and their families, and also help them to anticipate and prepare for the end of life. Multidisciplinary management in tertiary centers is recommended in close collaboration with general practitioners, home carers and a dedicated health network. Patients' follow-up deals mainly with motor impairment and physical disability, adaptation, nutrition and respiratory function. Involvement of palliative care as part of the multidisciplinary team management offers patients the possibility of discussing their end of life issues. This review summarizes the different aspects of ALS care, from delivering the diagnosis to the end of life, and the organization of its management.

1. Introduction

Amyotrophic lateral sclerosis (ALS) is a relentlessly progressive and fatal neurodegenerative disease. With an incidence of around 2/100,000 person-years, ALS is the most common form of motor neuron disease and the third most common neurodegenerative disease [1]. Degeneration of both the lower (LMNs) and upper motor neurons (UMNs) leads to progressive weakness of voluntary muscles of movement as well as the swallowing, speech and respiratory muscles. A variable degree of spasticity and pseudobulbar affect can be observed too. Roughly 50% of ALS patients develop mild cognitive and/or behavioral disorders and 5–15% develop dementia, usually of the frontotemporal type [2–4]. About 90% of ALS cases are sporadic of unknown origin, whereas the remaining 10% are classified as familial [5]. To date, the genetic etiology is identified in approximately two out of three cases of familial ALS and in around 10% of sporadic cases [6].

The presentation and progression of the disease as well as the age of onset vary considerably between individuals. The rate of disease increases with age to peak in the group aged 65–75 years. Nevertheless, disease can also arise in young adults (< 30 years) and in the elderly (> 80 years). Median survival time is 3–5 years from symptom onset, although 5–10% of patients may survive for a decade or more [1,7]. These data emphasize the necessary adaptation of clinical management according to the patients' age and course of disease.

The first consensus meeting on ALS management in France was held in 2005. More recently, French ALS care recommendations were reviewed as part of the French national protocol for diagnosis and care [8]. At the same time, a European

* *Corresponding author.*
E-mail address: soriani.mh@chu-nice.fr (M.-H. Soriani).
http://dx.doi.org/10.1016/j.neurol.2017.03.031

Federation of Neurological Sciences (EFNS) task force on the management of ALS and the American Academy of Neurology (AAN) published guidelines for clinical care in ALS [9,10]. In the absence of curative treatment, the aim of management is to maximize quality of life (QOL), prolong survival, and support ALS patients, their families and their caregivers to help them anticipate and prepare for the end of life.

The present review summarizes the organization of such management and the different aspects of ALS care, from delivering the diagnosis to the end of life, and includes neuroprotective and symptomatic treatments, nutritional and respiratory management, rehabilitation, psychological and social management and, finally, palliative and end-of-life care.

2. ALS centers and multidisciplinary management

Because of (i) the diversity of ALS damage, involving physical functions, breathing, nutrition, communication, cognition and emotions, in association with complex needs related to health and psychosocial aspects, and (ii) the progressive evolution of the disease with often rapid changes, multidisciplinary care (MDC) is recommended at tertiary centers (ALS centers) [9,11,12].

Patients receiving care at ALS centers have longer survival than those supported by general neurology clinics [12,13]. Most observational studies have shown that MDC is effective in several ways including increased use of (i) riluzole, (ii) non-invasive ventilation (NIV), (iii) adaptive equipment and, in some studies, increased use of gastrostomy [13]. Although certain treatments and assistive devices may increase survival, no study has yet clearly demonstrated their survival benefits for patients managed at ALS centers. However, better decision-making in complex situations by experienced practitioners may also contribute to better survival. In addition to extending survival, studies have demonstrated improved QOL for patients receiving care at ALS centers [14,15], with patients at ALS centers having fewer unplanned admissions for acute events, which contributes to the increased QOL for patients while decreasing the costs to the healthcare system, with no negative effects on patient outcomes [12].

The multidisciplinary ALS team usually includes healthcare practitioners and a social counsellor, and provides management services for ALS patients (Fig. 1). Tertiary centers offer appointments covered by a single visit, decreasing patients' trips to hospital and, consequently, their tiredness. Quarterly appointments are usually proposed, with variations according to disease progression. To facilitate patients' management, effective communication and good coordination are essential between the center's multidisciplinary team, home practitioners and the palliative care team.

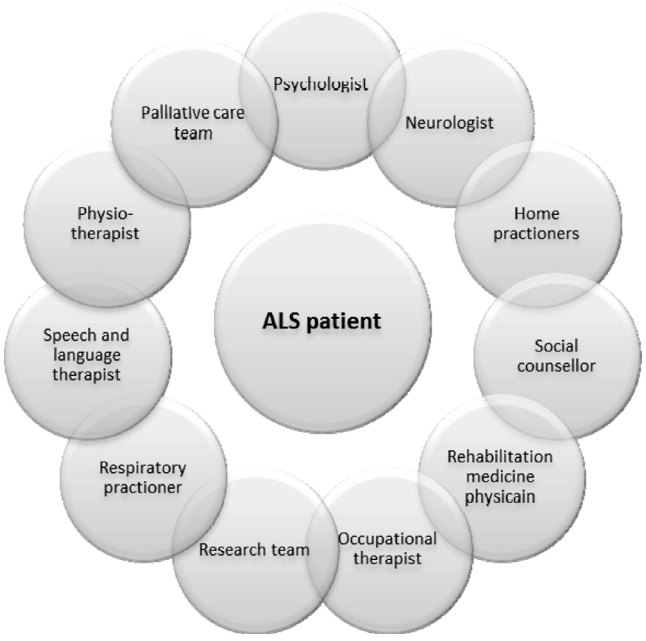

Fig. 1 – The multidisciplinary network of care in amyotrophic lateral sclerosis (ALS) tertiary care centers.

3. Breaking the news

Delivering the ALS diagnosis to patients and their relatives is challenging for both patients and clinicians. While there is no standardized procedure, it has been reported that many factors influence how the announcement of bad news is experienced [16–18]. Disclosing the diagnosis of ALS requires a specific environment. The physician should meet with the patient in a quiet comfortable room, with the presence of relatives if expected by the patient, as soon as the diagnosis has been confirmed. To help personalize the announcement, the physician should take into account the patient's personal history, social and environmental context, what the patient already knows about the disease and what he wants to know.

Such an announcement requires good communication skills, empathy, honesty, and use of simple and careful words. Information should include the name of the disease, the nature of its progression and the wide variability of the prognosis. Reassurance is important at this time and should comprise information about neuroprotective treatments and the research, and assure the patient and family that they will not be left on their own to face the disease. Time should be made available for the patient to ask questions. As diagnostic announcements may lead to psychological trauma, the intervention of a psychologist is usually proposed.

The patient's general practitioner should also be promptly informed by being sent a report specifying the delivered information.

Finally, to evaluate the patient's perception and understanding of the diagnosis, another appointment should be proposed in the month following the diagnosis. These two-tiered appointments give the patient the opportunity to handle the changed perspective of his life [19].

4. Therapeutic management

This is based on the establishment of neuroprotective and symptomatic treatments.