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Motor neuron diseases

Ethical considerations and palliative care in patients with amyotrophic lateral sclerosis: A review

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ABSTRACT

Amyotrophic lateral sclerosis (ALS) is not a curable disease, but it is treatable. By definition, much of the care provided to ALS patients is palliative, even though active life-sustaining strategies are available to prolong survival. Healthcare professionals must develop communication skills that help patients cope with the inexorable progression of the disease and the inevitability of death. Symptomatic treatments as well as respiratory insufficiency and nutritional life-sustaining therapies must be regularly evaluated as the disease progresses, without losing sight of the burden placed on the patient's non-professional caregivers. The decision-making process regarding tracheostomy with invasive ventilation (TIV) is of greater complexity. Providing full information is crucial. Several long interviews are necessary to explain, discuss and allow assimilation of the information. Also, physicians should be careful not to focus exclusively on the biomedical aspects of disease, as ALS patients generally welcome the opportunity to discuss end-of-life issues with their physicians. Psychological factors, education level and cognitive status (especially the level of executive dysfunction) have a major influence on their decisions. However, as many patients do not complete advance directives with regard to TIV, advance care planning may instead be suggested in anticipation of emergency interventions. This should be discussed by healthcare professionals and the patient, and based on the wishes of the patient and caregiver(s), and communicated to all healthcare professionals. Many healthcare professionals are involved in the management of an ALS patient: they include not only those at ALS centers who provide diagnosis, follow-up and treatment initiation (particularly for respiratory and nutritional care), but also the medical and social care networks involved in disability support

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and home care. Specialist palliative care teams can work in partnership with ALS centers early in the course of the disease, with the center coordinating information-sharing and collaborative discussions.

1. Ethical challenges

Amyotrophic lateral sclerosis (ALS) is a progressive fatal, incurable, disease. The prognosis is based on the patient's nutritional and respiratory status. The main goal of treatment in caring for ALS patients is to minimize morbidity and maximize quality of life (QoL).

1.1. *Medical information*

The provision of medical information on ALS often comes as an existential shock to the patient: it reveals the patient's powerlessness, finiteness and vulnerability, and destroys the image of omnipotent medicine that can always cure or, at least, repair. Healthcare professionals need to develop communication skills that help these patients manage the inexorable progression of the disease and the inevitability of death [for example, training in giving bad news, which is in line with the guidelines and procedures issued by health authorities, including the French National Health Authority (*Haute Autorité de Santé*)]. Indeed, healthcare professionals themselves may need psychological support or ethics resources to deal with their own emotional burden associated with delivering bad news.

1.1.1. *With severe forms of disease, the bad news keeps coming*

With a view to increasing survival time, therapeutic strategies should be rapidly implemented to compensate for malnutrition and respiratory insufficiency (sleep disorders, in particular) and to prevent aspiration, falls and pneumonia [1]. However, transient psychological reactions (such as denial and shock) can delay treatment initiation.

These treatments are of major importance: early non-invasive ventilation (NIV) can improve QoL and increase survival time by >12 months [2]. A 5% weight loss (relative to the patient's usual body weight) at diagnosis multiplies the risk of death by a factor of two, whereas a gain of more than 2.5 kg in fat mass is associated with a 10% reduction in the risk of death [3]. Conversely, at more advanced stages of the disease, declining respiratory function means that the use of gastrostomy for nutritional support is probably of little benefit and may even be associated with increased mortality [4].

The manner in which information is delivered to ALS patients is acknowledged as the first important step in their care [5]. It is difficult to announce the need for gastrostomy or NIV to the patient because both events are related to life expectancy and burdensome treatment procedures. One prospective study has highlighted the complexity involved in a patient's acceptance of NIV and gastrostomy: in addition to education level and cognitive status (especially the level of executive dysfunction), psychological factors also have a major influence [6]. Studies have found that patients with psychological distress (such as stress, depression, hopelessness) have a greater risk of dying than patients with psychological well-being [7].

1.1.2. *People differ in the speed with which they take in bad news*

According to French legislation (the 2002 Patients' Rights Act and article 1111-2 of the French Public Health Code), the physician must respect a patient's right not to be given information that may be unbearable or shocking: ''A person's wish not to be informed of a diagnosis or a prognosis must be complied with, except when there is a risk of transmission to a third party.'' The process is difficult, and physicians need to take into account the psychosocial and existential effects of ALS: time should be allowed for explanation, assimilation and discussion; and several long interviews with the physician are required to ensure that the information has been understood [8].

In addition, the manner in which the physician speaks to the patient is more important than the content: ''the important thing is to tell the patient what he/she is capable of hearing and to tell him/her in such a way that he/she can be told more later on'' [9]. To this end, the information should not be passively received by the patient, but should be the result of a shared process of interaction. Patients need to be told about the disease (the diagnosis, about which they as yet know nothing) on top of what they already know; telling people they are ill amounts to reaffirming the vulnerability of the human condition and the finiteness of our existence. A person only really becomes a patient once the diagnosis has been announced. According to Delassus, this announcement can trigger a sudden awareness, a breakdown, ''chaos'', a ''terrible overdose of knowledge'', which can interfere with the patient's care and activities of daily living (ADL). Such an announcement may also often lead to denial or shock, which can overwhelm and 'paralyze' the patient. Accordingly, Delassus emphasizes the need for (i) cautious information delivery and (ii) awareness of the effects that announcing the diagnosis may have. Good practice guidelines for neurologists on how best to do this are available (Table 1) [10,11].

Some researchers offer advice on how to communicate bad news in an ethical way, which touches on issues of autonomy and responsibility [12]. Qualitative studies of patients' satisfaction regarding the way the diagnosis was communicated have highlighted the importance of exploring how much information patients want to receive and what they are