

# Management of patients with amyotrophic lateral sclerosis

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## Summary

Standard recommendations for the clinical management of patient with ALS have been edited in recent years. These documents emphasise the importance of patient's autonomy.

*Aim of study:* To measure how these different recommendations can be applied in the context of a general hospital without a specific ALS clinic.

*Methods:* Review of medical records of 21 patients with an ALS diagnosis treated by the University Hospitals Geneva who died from 1996–2002.

*Results:* Patients suffered from distressing symptoms during their last hospitalisation. Artifi-

cial nutrition was given to 5 patients. Six patients had non invasive ventilation (NIV). Written advance directives were only available in 2 cases. Discussions about these issues were also conducted late in the evolution of the disease.

*Conclusion:* Some discrepancies between our daily practice and the existing recommendations exist, particularly regarding the key issues of artificial nutrition and ventilatory support.

*Key words:* amyotrophic lateral sclerosis; palliative care; advance care planning

## Introduction

Amyotrophic lateral sclerosis is the most common degenerative motor neuron disorder occurring in adult life. Its prevalence has been estimated at 6–8/10<sup>5</sup>, with an incidence of 2/10<sup>5</sup> per year [1]. To date, no curative treatment exists for ALS and symptomatic care must be provided ab initio [2–4].

Recommendations for the clinical management of patients with ALS and their families have

been reviewed in recent years by a subcommittee of the American Academy of Neurology (AAN ALS practice parameters) [5].

The aim of the study was to better characterise the end-of-life of patients with ALS and to measure how these different recommendations could be applied in the absence of a specific ALS clinic.

## Methods

This study is a review of medical records of patients with a diagnosis of ALS hospitalised at the University Hospitals of Geneva, who died during the 1996–2002 period. According to the AAN ALS practice parameters, information concerning the introduction of artificial nutri-

tion or ventilatory support, as well as the completion of advance directives were noted. Main symptoms present during the last hospitalisation, symptom management and circumstances of death were also collected.

## Results

Twenty-one (12 women, 9 men) patients were included in the study. Median age of the patients was 64.2 years (58–80). Twenty patients lived at

home and one in a nursing home. Twelve patients were hospitalised at least once in the year before death. Ten patients died in the geriatric ward (4 pa-

tients in a palliative care unit), 6 in the long term care ward, 2 in the acute care ward, 2 at home and 1 in a nursing home. Median length of the last hospitalisation was 42 days. Median length of evolution of ALS from diagnosis until death was 20.8 months (range: 6–60). Reasons for the last admission were general weakness (n = 10), anxiety and/or depression (n = 8), pulmonary infection (n = 3), dysphagia (n = 3) and exacerbation of dyspnoea (n = 2). Comparison of the recommendations of the AAN ALS practice parameters and our findings are listed in table 1.

### Symptom management and palliative care

All patients had some degree of communication impairment and general weakness. Eleven patients had constipation, 8 had insomnia and/or anxiety. Sixteen patients were treated with riluzole until their last days of life; 6 patients received antibiotics for pulmonary infection.

### Nutrition

Although all patients suffered from dysphagia and significant weight loss (>20% of body weight prior to diagnosis), only 5 patients had received a percutaneous endoscopic gastrostomy (PEG), and one patient had parenteral nutrition. Artificial nutrition was introduced 7 months (mean, range: 2–13) before death. Two patients, hospitalised for PEG installation, died during the hospitalisation. Eight patients refused PEG before or during hospitalisation. Nutrition was continued until the last day for all patients with artificial nutrition.

### Respiration

To alleviate symptoms of chronic hypoventilation, 6 patients had non-invasive ventilation (NIV) with a nasal mask. Median length of NIV was 9.5 months (2–24). For 2 of these patients, NIV was introduced during an emergency hospitalisation for bronchopneumonia. No patient had a tracheotomy. No patient asked to stop NIV before his death. Among the 15 other patients, 3 declined

**Table 1**

Comparison of AAN ALS practice parameters (left column) and our findings (right column).

#### Breaking the news:

The physician should give the diagnosis to the patient and discuss the implications.

The diagnosis should always be given in person and never by telephone.

Provide printed materials about the disease and about support and advocacy organisations, and a letter or audiotape summarising what the physician has discussed.

Avoid the following: withholding the diagnosis, providing insufficient information, delivering information callously, or taking away or not providing hope.

Data not collected in our study

#### Symptomatic management:

##### *Sialorrhoea:*

Treat sialorrhoea with glycopyrrolate, benzotropine, transdermal hyoscine, atropine, trihexyphenidyl hydrochloride, or amitriptyline.

Treat thick mucus production associated with sialorrhoea with propranolol or metoprolol.

Consider manually assisting coughing and mechanical insufflation-exsufflation for clearing secretions, especially during acute infection.

8 patients received glycopyrrolate associated with respiratory therapy.

##### *Pseudobulbar affect:*

Treat pseudobulbar affect with amitriptyline.

Consider fluvoxamine as an alternate choice.

No pseudobulbar effect was noted. 7 patients received antidepressants (SSRI) for depressive mood

#### Nutrition:

PEG is indicated for patients with ALS who have symptomatic dysphagia and should be considered soon after symptom onset.

For optimal safety and efficacy, PEG should be offered and placed when the patient's VC is more than 50% of predicted.

5 patients had a PEG tube introduced 7 months (mean) before death.

#### Respiratory management:

Be vigilant for symptoms indicating hypoventilation. Offer noninvasive ventilatory support as an effective initial therapy for symptomatic chronic hypoventilation and to prolong survival in patients with ALS

When long-term survival is the goal, offer invasive ventilation and fully inform patient and family of burdens and benefits.

In accordance with the principles of patient autonomy, physicians should respect the right of the patient with ALS to refuse or withdraw any treatment, including mechanical ventilation.

When withdrawing ventilation, use adequate opiates and anxiolytics to relieve dyspnoea and anxiety.

6 patients were using NIV until their last day of life.

No invasive ventilation was used.

#### Palliative care:

##### *For pain management:*

Use non-narcotic analgesics, anti-inflammatory drugs, and antispasticity agents for initial treatment of pain in patients with ALS., and opioids for patients who do not respond to the above.

##### *For treating dyspnoea in terminal stages of ALS:*

Use opioids alone or with supplemental oxygen, to treat dyspnoea at rest in patients with ALS, despite the risk of respiratory depression with higher doses.

15 patients had pain, 12 received morphine, 1 paracetamol and 1 anti-inflammatory drugs.

18 patients received supplemental oxygen and 12 patients received morphine.

#### For hospice care:

Consider referral to hospice in the terminal phase of ALS.

4 patients died in a palliative care unit. 5 patients had pain and palliative consultation

#### For advance directives:

Initiate a discussion of advance directives well in advance of the terminal phase and re-evaluate at least every 6 months.

2 patients had written their advance directives shortly before death.

NIV, 2 patients couldn't tolerate the mask, one patient had no medical indication for NIV and for one patient NIV was discussed too late in the evolution of the disease.

#### Advance directives

Only two patients had written their advance directives. In both cases this was done during their last hospitalisation, shortly before death.

#### Circumstances of death

Nine patients died of pulmonary infection, 7 patients became comatose and died and 3 patients were found dead in their bed. Three patients with refractory dyspnoea were given terminal sedation with midazolam [6].

## Discussion

Most of the patients had active symptom management for several distressing symptoms in the last weeks or months of their life, as suggested in the recommendations. Furthermore patients presented with different symptoms that are not addressed in the actual recommendations like communication impairment, general weakness, insomnia, anxiety or constipation [7]. Other questions are not raised in the recommendations such as when to stop riluzole and when to introduce antibiotics. Our results demonstrated some discrepancies between our daily practice and the existing recommendations in the key issues of artificial nutrition and ventilation. These results are similar to Bradley's study that measured the effect of the AAN ALS practice parameters on the management of ALS. Though the pharmacological symptom management improved significantly, many patients did not receive artificial nutrition or ventilatory support, mainly because lack of patient compliance [8]. In our study, only 6 out of 21 patients (29%) had a PEG performed, which is in the lower range of previously reported studies [7, 9, 10]. Different reasons can be hypothesised to explain this low rate. Patients with ALS do not want their life to be prolonged, since it has been suggested that the early use of PEG may prolong survival, even if no randomised trials are available [5, 11–13]. Another reason for refusing artificial nutrition might be that patients want to eat naturally until the end of their life. Finally, this issue may have been discussed with the patients too late in the evolution of the disease, or not discussed at all. Non invasive ventilation (NIV) via a facial or nasal mask is an effective way of alleviating symptoms of chronic hypoventilation and respiratory muscle failure such as daytime sleepiness, dyspnoea, orthopnoea, sleep disruption, morning headaches

and cognitive disorders [4]. In spite of this, although 83% of patients had dyspnoea, less than a third of the patients included in our study had NIV. These results are comparable with other studies, suggesting an under-use of ventilatory support in symptomatic patients [7, 9, 10]. Only two patients in the present study were unable to tolerate the mask or could not use the device. In our study NIV was not proposed systematically to all patients because either the discussion about NIV occurred too late in the evolution of the disease or patients did not want their life to be prolonged [14, 15]. No patient had either a tracheostomy or invasive ventilation. As in several other European countries, invasive ventilation via tracheostomy is an option that is actively discouraged in ALS patients in Switzerland [16].

In 1996, in the Canton of Geneva, a specific legislation relative to advanced directives has been implemented [17]. However, only 2 of 21 patients included in our study had written advanced directives, thus, suggesting lack of information among patients and health professionals in our area. In conclusion, even in lack of ALS clinic, we would recommend the implementation of a multidisciplinary consultation for patients with ALS to support the completion of advance directives and early discussion about artificial nutrition and ventilatory support.

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