Société Française de Neurologie

Association des Neurologues Libéraux de Langue Française

Consensus conference
guidelines

Care of patients with amyotrophic lateral sclerosis (ALS)

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QUESTIONS PUT TO THE JURY

Question 1. How to diagnose ALS?

Question 2. How to break the news of ALS?

Question 3. How to assess the course of ALS and what instruments to use?

Question 4. What therapies and follow-up are available for patients with ALS and their families?

Question 5. What is the role of life support in patients with ALS?

Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disease affecting both upper and lower motor neurons of the voluntary motor pathway. It is fatal in the medium term as there is no cure. Its peak incidence is around the age of 50. It manifests as progressive motor weakness affecting the limbs and the labial and glossopharyngeal muscles. These deficits lead to loss of autonomy and require early, continuing and graduated multidisciplinary management so that the patient can stay at home. However, because the disease is severe and often prolonged, new ways of organising care need to be found collectively and implemented, with special emphasis on the needs of the patient, carers and family, and on interdisciplinary teamwork.

QUESTION 1. How to diagnose ALS?

- ***When is ALS suspected?***

ALS should be suspected when there is a mixed lower motor neuron (LMN) and upper motor neuron (UMN) deficit and progressive spread of symptoms. Diagnosis is based on clinical examination and electrophysiological (electromyography and nerve conduction velocity studies) performed by a neurologist.

- *LMN deficit* is diagnosed from:
  - clinical data: motor weakness with amyotrophy and fasciculations spreading progressively, but with preservation of ocular motility, objective sensory function and sphincter function;
  - electrophysiological studies: signs of denervation affecting the bulbar, cervical and lumbar muscles, but with no motor conduction block on the roots or peripheral nerves and no neuromuscular junction disorders.
- *UMN deficit* is diagnosed mainly from clinical data: exaggerated reflexes in a patient with muscle weakness and amyotrophy.
- There is no inflammatory syndrome in the blood or cerebrospinal fluid.
- Magnetic resonance imaging shows no motor nerve root or spinal cord compression.

- ***Differential diagnosis***

The possibility of other motor deficit diseases (eg myasthenia, motor conduction block neuropathy, cervical syringomyelia) should be eliminated as they can resemble ALS in its early stages.

- ***Genetic tests***

Genetic tests should be ordered only in cases of familial ALS (at least two family members affected irrespective of degree of relationship) or if the phenotype is atypical.

QUESTION 2. How to break the news of ALS?

Telling a patient they have ALS and explaining the course of the disease may cause major repeated psychological trauma which the patient will have to cope with (defence and adaptation mechanisms). Clinical management of ALS should include knowledge, understanding and acceptance of these mechanisms. Support from a clinical psychologist is usually required.

Breaking the news is an interactive dialogue where providing medical information and listening to the patient are equally important. Whenever possible, the neurologist who will be caring for the patient should break the news.

Throughout the course of the disease, a balance should be maintained between anticipating onset of handicap and introducing life support measures, on the one hand, and respecting the patient's psychological state, life plans and environment, on the other.

QUESTION 3. How to assess the course of ALS and what instruments to use?

- ***Regular assessment***

Because ALS is a progressive disease, the patient's condition, their needs, and those of carers should be assessed regularly, if possible within the home. By assessing deficits and how they affect what the patient can do, the course of the handicap and deterioration in vital functions can be anticipated. For maximum efficacy, assessments should be coordinated by ALS centres that can provide a multidisciplinary consultation at a single site.

- ***Initial tests, repeated every three months:***
  - interview: situation with regard to family, work, social life, home environment, etc.
  - disability: ALS Functional Rating Scale (ALSFRS-R), (functional independence measure (FIM), Barthel index)), walking range, pain
  - manual muscle testing
  - speech therapy assessment: dysarthria, swallowing, communication
  - psychological assessment
  - nutritional status: weight, height, body mass index (BMI), dietary habits
  - respiratory function:
    - signs of sleep disorders, dyspnoea, orthopnoea, bronchial congestion,
    - vital capacity, peak expiratory flow (PEF) with cough test, maximal inspiratory pressure (PI max), sniff nasal inspiratory pressure (SNIP), nocturnal oximetry (every 6 months), blood gas analysis (at the outset, then according to disease progression).

QUESTION 4. What therapies and follow-up are available for patients with ALS and their families?

- ***Drugs for ALS***

Riluzole (100 mg/day) is the only active drug with a marketing authorisation for ALS. It should be prescribed as soon as a diagnosis of ALS is suspected.

- ***Management by a multidisciplinary team***

Management should be coordinated, flexible, and adjusted for the patient's stage of deficit by the professional team (physiotherapist, occupational therapist, speech therapist, nurse, nursing auxiliary, home help, medical and personal support assistant, social worker, psychologist, dietician, beautician, carers (family, volunteers), service providers). Each team member helps the patient cope as well as possible with their handicap under the conditions of greatest comfort.

- ***Symptomatic drug therapies***

— *Non-specific symptoms* (fatigue, depression and anxiety, constipation, pain, cramp). There are no ALS-specific drugs for these symptoms. Conventional treatment should be given in accordance with good clinical practice.

— *Specific symptoms or symptoms requiring special treatment because they have occurred in a patient with ALS.*
  - Salivary disorders:
    - stasis: tricyclics, scopolamine, botulin toxin A or radiotherapy
    - change in consistency of saliva: beta-blockers
    - night-time dry mouth combined with daytime stasis: difficult to treat.
  - Spasticity: baclofen, tizanidine, dantrolene or benzodiazepines.
  - Emotional lability: tricyclics or serotonin reuptake inhibitors.
  - Sleep disorders: look for respiratory problems and for anxiety and/or depression:
    - antihistamines or hypnotics; avoid benzodiazepine derivatives if possible.

- ***ALS centres***

ALS centres have greatly improved patient care by providing a formal framework. A local coordinator arranges all aspects of interdisciplinary care. The patient record remains the patient's property and is shared by all those involved.

QUESTION 5. What is the role of life support in a patient with ALS?

- ***Arriving at a joint decision***

Because ALS is ultimately a fatal disease, a joint decision about possible nutritional and/or respiratory assistance should be discussed by all parties fairly early in the course of disease, well before it is needed. The aim would be to include the patient in the decision-making process, in a climate of trust and in a medical, psychological and social situation prepared for the consequences of permanent life support at home.

- ***Palliative care***

Palliative care should be discussed well before it is likely to be needed if the patient seems willing to do so. The patient's wishes should be complied with in this respect. Whatever the patient's decision early on during the course of the disease, they should be able to request and benefit from palliative care at any time.

- ***Enteral feeding***

Enteral feeding prevents the effects of malnutrition, improves patient comfort and may prolong survival. It should be given via a percutaneous endoscopic gastrostomy tube. The clinical criteria for starting enteral feeding are feeding problems.

- ***Respiratory assistance***

Respiratory assistance - either non-invasive ventilation (NIV) using a mask or tracheotomy - should be given at home. It is clear that it improves comfort, sleep and survival, but it does not stop disease progression. It should be started when the patient has symptoms of alveolar hypoventilation. The patient and carers should be aware that dependence on respiratory assistance will gradually increase.

- ***Acute respiratory failure***

In the event of acute respiratory failure, the decision to provide ventilation (NIV or intubation), palliative care, or sedation during the terminal stage should be taken after considering factors in the patient's history, any wishes expressed by the patient, his or her personal and family history, and any uncertainty about whether or not the acute respiratory failure is reversible. When deciding on the best form of care, the patient's comfort and support for the patient and carers are taken into consideration.

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The long version of the guidelines may be obtained (in French) by writing to: Haute Autorité de santé Service communication, 2, avenue du Stade de France – 93218 Saint-Denis La Plaine CEDEX France. It is also available on the HAS website www.has-sante.fr under "Publications".