RESPIRATORY FAILURE IN PATIENTS WITH AMYOTROPHIC LATERAL SCLEROSIS (ALS): POSSIBILITIES AND LIMITS

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SUMMARY

The Authors aim to assess the possibilities and limitations of the respiratory treatment of patients with amyotrophic lateral sclerosis. After having said that the goal of treatment is to slow the course of the disease they took on the pharmacological treatment support with antioxidants in combination with drug therapy and conclude by analyzing the different types of ventilation used in various forms because of the severity of respiratory failure.

Key words: Amyotrophic lateral sclerosis; motor deficit; spasticity; antioxidant treatment; mechanical ventilation

RIASSUNTO

Gli Autori si propongono di valutare le possibilità e i limiti del trattamento dell’insufficienza respiratoria dei pazienti con sclerosi laterale amiotrofica. Dopo avere premesso che l’obiettivo del trattamento è quello di rallentare il decorso della malattia si soffermano sul trattamento farmacologico di supporto con sostanze antiossidanti in associazione alla terapia farmacologica e concludono analizzando i vari tipi di ventilazione utilizzabili nelle varie forme di graviità dell’insufficienza respiratoria.

Parole chiave: Sclerosi Laterale Amiotrofica, deficit motorio, spasticità, trattamento antiossidante, ventilazione meccanica

Introduction

The amyotrophic lateral sclerosis is a disease with sporadic autosomal-dominant transmission who prefers adulthood – elderly age (with a peak between 50 and 70 years) and that only in 10% of cases is hereditary (SLAF, amyotrophic lateral sclerosis family, which is transmitted by an autosomal recessive trait), and in this case the age of onset is lower (around 40 years).

The disease has a progressive worsening course and usually leads to death (in the absence of controlled ventilation) in 2-3 years for various complications (frequent aspiration pneumonia due to dysphagia from involvement of cranial nerves IX-X, for uncoordinated pharynx-esophageal muscle).

Recent statistics, however, showed that 50% of ALS patients survive beyond 3 years after diagnosis and 10% after 10 years. The paralysis of respiratory muscles needed assisted ventilation and is unfortunately a frequent cause of death for people with ALS.

Diagnostic criteria

The diagnosis is based on clinical criteria supported by instrumental examinations: chest x-ray, spirometry (to assess the vital capacity VC and Maximum Inspiratory Pressure MIP) emogasanalysis to assess ossiemia and capnia, oximeter night bacteriological spontaneous and/or induced sputum examination, bacteriological examination of bronchoaspiration or brushing (periodic inspection every three months).

The EMG dependent muscles involved shows signs of suffering of the second motor

Therapeutic interventions

There is not now an official therapy for ALS. The goals of treatment are to slow the course of the disease and improve the quality of life of the patient through the monitoring of the most debilitating symptoms and correction of motor deficits, communication and swallowing.
As for drug therapy currently have of Rilutek (riluzole), the supportive care is the administration of antioxidants such as vit E, reduced glutathione and creatine monohydrate and the symptomatic therapy involves the use of muscle relaxants for spasticity and cramps (es. Lioresal and Sirdalud), anticholinergics for the excessive salivation, antidepressants for depressive reaction, non-benzodiazepine hypnotics for sleep (zolpidem, zopiclone) and non-benzodiazepine anxiolytic for anxiety (buspirone). To avoid, in the presence of signs of respiratory failure, the administration of benzodiazepines (like Valium or Control) to sleep or anxiety and expectorant (Fluimucil, Fluifort) that may increase secretions. The symptoms of chronic nocturnal hypoventilation can seriously deteriorate the quality of life of patients. The intermittent non invasive ventilation with mask (NIV) is an effective and convenient way to alleviate these symptoms and may even prolong considerably the life expectancy of patients.

Possible therapeutic interventions, however, are different according to the degree of impairment: in a first stage will be sufficient breath physiotherapy, abstaining from smoking, weight loss if you are overweight and avoid fatigue. When obstruction of airway predominate will be useful pharmacological prevention of secretions and infections with low doses of cortisone and antibiotics in cycles and aspiration of secretions using a suction mechanic, a small tube that is placed behind the tongue and connected to an engine that performs the aspiration.

The patient with ALS who has moderate breathing difficulties or at night may benefit from nocturnal nasal ventilation. In this cases we can use of mechanical ventilators that breathed air directly into the airways through a mask placed in front of the mouth. Bilevel PAP is used when the respiratory disorder is predominantly obstructive type and IPPV prevails if there is failure of respiratory muscles. All these ventilators are portable and easy to manage at home.

When the breathing difficulties are severe these supports are insufficient and so Tracheostomy becomes necessary. Tracheostomy is a very simple and painless allowing you to place an endotracheal tube that is connected to the ventilator.

You can resort to invasive ventilation in certain circumstances, for example, when the bulbar deficit make it impossible for the non-invasive ventilation, and when you want to accept a permanent ventilation support.

Discussion

Respiratory complications are due to 3 main mechanisms: the weakness of respiratory muscles, ineffective cough with accumulation of secretions and bulbar paralysis. These complications mostly occur late, but in some cases may arise even in the early stages of the disease. It is important to undergo regularly, at least every 6 months, to monitoring tests of respiratory function.

Factors such as a slow progression, good communication skills, a mild involvement of bulbar and a strong motivation by the patient and a family collaborative atmosphere, promote the start of NIV. It is very important to reassure the patient that if he/she decides to stop the NIV, will be available all the necessary care and appropriate drugs to prevent death by suffocation. The liaison with the hospital and extra-hospital (hospice) structure can give great help in these cases.

If the patient refuses to take the NIV we can use the application of intermittent oxygen. The start of the respiratory failure is hardly an acute event and in most cases manifests itself subtly often with symptoms that are not as important as the headache on awakening, daytime sleepiness, the feeling of “lack of air” after an effort and tachycardia.

If assistance is complete, people with ALS who use mechanical ventilation and all other necessary treatments may live for many years. However there are problems that arise with the use of invasive ventilation in the long term:

• the quality of life of the patient may fall sharply leading to a low view of themselves;
• 5-10% of people with ALS develop a state of locked-in, meaning that they are unable to communicate in any way;
• there is the possibility of having to resort to a long period of hospitalization;
• the lives of family members can become demotivating and fragmented, leading to states of tension, anger and feelings of imprisonment;
• personal financial resources and family can be reduced.

The indications for ventilation with a tracheotomy include a reduced control of symptoms or a persistent high level of CO₂, despite the use of a non-invasive ventilation, ineffective cough and inability to rid the bronchial secretions, suction problems.

This type of ventilation has several disadvantages. Some people have the sensation of a foreign
body, which increases their disability and dependency. Secretions requiring suction, either at night or during the day. Cough to liberate the small airways is difficult. The seat of the tracheotomy can cause infection, bleeding and could produce an inflamed tissue (granuloma) that requires removal. It requires more practice and more attention, and is more complex to manage. Furthermore, some people have problems swallowing and language after tracheotomy.

The unwanted intubation performed by emergency physicians in the face of terminal respiratory failure is not uncommon when the patient and family are poorly informed about the disease. Such patients can survive for years within the intensive care unit and move towards a complete "deafferentation syndrome " (locked-in syndrome) when any communication with the outside world is rendered impossible by a complete tetraplegia which also involves the extraocular muscles.

Conclusions

The desire to stop using life support in these cases brings medical, ethical and legal problems. Therefore, an important goal of a good doctor-patient relationship in ALS must be to clearly and early discuss on the advantages and disadvantages of these processes and the development of advance directives that include an agreement on the conditions of interruption of each ventilatory support when there is no longer possible.

The decision to undergo Tracheostomy and artificial ventilation should be taken only by the patient and persons who assist him when the urgency of action is not yet expressed.

It is the doctor who establishes the need for data on the basis of clinical and instrumental in its possession, but it is the patient who decides whether he wants it or not, knowing clearly the prognosis of the disease and the problems and benefits that this choice entails.

References


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