CARE FOR PATIENTS WITH AMYOTROPHIC LATERAL SCLEROSIS – ADVANCED PRACTICES OF THE LAST DECADE

Definition and short description

Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disease, characterized by loss of motor neurons in the spinal cord, brainstem and motor cortex. The cause for ALS is still unknown, but there are many successful ways so far to maintain the condition and quality of life of the patients.

Medicinal treatment

Currently, there are no medications that can lead to a cure, but during numerous double-blind and placebo-controlled trials the best results were given by the drug riluzole regardless of dosing regimen. Hopes for improving the survival by lithium carbonate are not justified, as well as the dietary addition of antioxidants, vitamin E, acetylcysteine, selenium, l-methionine and creatine.

According to various authors, there is a clear evidence of better survival and quality of life of ALS patients when treatment is performed by a multidisciplinary team in specialized medical institutions. It must include neurologist, pulmonologist, physical and rehabilitation medicine specialist, speech therapist, psychologist, nutritionist and social worker. In cases, where it is impossible to form such a team, the telemedicine could be used to make the necessary consultations.

Clinical manifestations and most effective methods to deal with them

- One of the main difficulties for patients is the coming dysphagia, which leads to dehydration and weight loss. Although accurate signs for it are not formed yet, the percutaneous endoscopic endostomy may be regarded as a reliable way to correct this problem, as well as adding to the diet of nutritional supplements such as carnitine - 5-10 g per day, vitamin E - 5000 mg per day. However, if these supplements are not combined with riluzole, their effect is unknown;
- Respiratory failure is a leading cause of the worsening survival in ALS patients. Non-invasive ventilatory therapy (NVT) is discussed as a factor in improving the quality of life of patients. The main criteria for NVT inclusion are the nocturnal oximetry and bulbar involvement. The weakness of expiratory muscles leads to ineffective coughing, mucus retention in the upper respiratory tract and lung infections. Appropriate methods to assist expectoration are the mechanically-aided inspirium and expirium and high-frequency vibration on the chest wall;
- Sialorrhea – amitriptyline, botulinum toxin type B, injected into the salivary glands, radiotherapy, administered once;
- Pseudobulbar affect – fixed dose combination of dextromethorphan/quinidine;
- Fatigue can sometimes be a side effect of riluzole, so reducing or stopping the medicament for a while can be considered. However, the most significant results come by regular exercises according to the patient's state and capacities;
- Cramps – no drug has proven effect on this symptom so far, although gabapentin, vitamin E and riluzole, as well as levetiracetam, vitamins of group B and calcium channel blockers have been applied;
- Spasticity – the effect of baclofen, dantrolene and tizanidine is still without convincing results, but they are applied in severe cases in combination with specialized kinesitherapy to reduce spasticity;
- Depression – most authors suggest including treatment for depressive syndrome in all ALS patients;
- Anxiety – there are not enough studies on this issue. Different methods of psychology and physical therapy have good prognosis;
- Insomnia – previous studies suggest that there is no specificity in ALS treatment here and schemes valid for insomnia in general can be applied too;
- Cognitive and behavioral disorders – multiple teams are working to improve the condition of ALS patients through specialized methods of psychology and pedagogy;

- Dysarthria and other difficulties in communication – various options to improve speech abilities are discussed. There are devices for voice amplification, augmentative alternative communication, prosthetics, palate lifting and others.

Role of physical rehabilitation

In most patients supportive rehabilitation is a method of improving quality of life and periodic assessment of the condition in order to adequately include or exclude other medicinal and supportive therapeutic tools. Carrying it out in specialized institutions guarantees the quality of care and is a starting point for more detailed follow-up of the effectiveness of different therapies in this pathology. Combining preformed and natural physical factors implies more long-lasting results, but treatment programs must be adapted to the individual needs of each patient. Advanced approaches include the application of electrotherapeutic procedures, thermal and physical therapy, focusing on current deficits. The aim is each subsequent course of therapy to include other factors with complementary effects and to redefine the repeating of them according to the individual tolerance. It is advisable to conduct 4-5 courses in outpatient settings per year and one to be dedicated to climate- and water-treatment in sanatorium conditions.

References

1. McCluskey L, Casarett D, Siderowf A. Breaking the news: a survey of ALS patients and their caregivers. Amyotroph Lateral Scler Other Motor Neuron Disord 2004; 5:131–135.
2. Numico G, Anfossi M, Bertelli G, et al. The process of truth disclosure: an assessment of the results of information during the diagnostic phase in patients with cancer. Ann Oncol 2009; 20:941–945
3. Chio A, Bottacchi E, Buffa C, Mutani R, Mora G. Positive effects of tertiary centres for amyotrophic lateral sclerosis on outcome and use of hospital facilities. J Neurol Neurosurg Psychiatry 2006; 77:948–950.
4. Sorenson EJ, Mandrekar J, Crum B, Stevens JC. Effect of referral bias on assessing survival in ALS. Neurology 2007; 68:600–602.
5. Jackson CE, Gronseth G, Rosenfeld J, et al. Randomized double-blind study of botulinum toxin type B for sialorrhea in ALS patients. Muscle Nerve 2008; 39:137–143.
6. Robinson KM, Lacey SC, Grugan P, Glosser G, Grossman M, McCluskey LF. Cognitive functioning in sporadic amyotrophic lateral sclerosis: a six month longitudinal study. J Neurol Neurosurg Psychiatry 2006; 77:668–670.

To get further information on ALS, as well as on opportunities for medical rehabilitation and patients' training, please contact Medical Centre "RareDis".

E-mail: medical@raredis.org
Postal address: 24, Landos Street, floor 1, 4000 Plovdiv, Bulgaria
Phone: (+359 32) 577 447
Internet site: www.medical.raredis.org