Summary

Amyotrophic lateral sclerosis (ALS) is a primordial degenerative disease of a nervous system. Both its aetiology as well as pathogenesis remain unknown. ALS is characterized by gradually developing death of neurons which results in the damage to both upper and lower motor neurons.

The disease advances dramatically and results in deterioration of patient's ability to move (progressive muscular atrophy), increasing difficulty in articulation (dysarthria) and degradation of the natural way of swallowing food and saliva ((dysphagia).

Almost all affected patients experience speech disorders (dysarthria) turning into anarthria as the disease progresses. The authors report that 80%-95% of patients who suffer from ALS are unable to cope with everyday communication problems using natural speech. Unfortunately, when intelligibility starts to decrease, speech performance deterioration process is so fast, that no time remains left to implement appropriate augmentative and alternative communication method. Hence, proper and instant recognition constitutes the core factor in making clinical decisions.

So far, no comprehensive studies have been undertaken to establish a pattern of the course of disorders of individual articulation functions in patients with amyotrophic lateral sclerosis and no attempts to systematize them. It is important and justifiable to take such measures, because their results will allow for the creation of standards of neurological therapy in the diagnosis and therapeutic process of ALS patients. This will translate directly into an improvement in the quality of life and the therapeutic process of these patients.

The doctoral thesis aimed at assessing the dynamics of disorders of articulatory functions in the course of the limb form of amyotrophic lateral sclerosis (ALS). Main focus was on determining the direction and actual rate of degradation of articulatory functions and showing the differences in the course of disorders of articulatory functions depending on patient's sex.

The reference group of patients consisted of adult males and females,, treated in the Department of Neurology and Neurosurgery at the University of Warmia and Mazury in Olsztyn (UWM).

Heterogenic group of 60 people. 29 females and 31 males, aged 29-63. The oldest female being 63 on the first day of the research, the youngest 30. The oldest male was 62, the youngest 29. The patients in different stages of the disease. The avarage ALS Functional Rating Scale in

the group was 34 (the highest value 48 and 20 as the lowest value). None was on the PEG feeding system. All treated with Riluzole.

The patients not receiving any treatment nor neurologopedic therapy before the onset of ALS, after the diagnosis and in the course of the disease.

Inclusion criteria for patient's participation in the study:

1. Age ranging between 18-70.
2. Diagnosed ALS, exclusive of bulbar onset and classified according to El Escorial criteria.

Exclusion criteria:

1. The study also excluded ALS patients with bulbar dysfunction due to the fact that in this form, degradation of articulation functions is so rapid, that it makes it impossible to capture the actual advance in disorders.
3. Escalation of symptoms – not less than 56pts in the Frenchay Dysarthria Assessment Scale

During the FDA assessment process a patient could receive a maximum of 112 points (28 assessed functions on a 5-point scale, where the maximum is 4 points and the minimum is 0 points. Only those who received 4 or 3 points for each of the assessed articulation functions in the initial part were included into the study. A smaller numer of points indicated too highly developed dysarthia symptoms and consequently, the degradation of articulation functions could not be assessed over the time of six months. Patients were examined three times in equal time spans, every three months:

- first initial examination
- second examination after three months
- third examination after six months

28 articulation functions were assessed, including eight primary ones: reflex(cough, swallow,dribble) respiration (at rest, in speech) lips (at rest, spread, seal, alternate, in speech) jaw (at rest, in speech) palate (fluids, maintenance, in speech) laryngeal (time, pitch, volume, prosody) tongue (at rest, protrusion, elevation, lateral, alternate, in speech) intelligibility (repetition, description, conversation).

The results of tests assessing the dynamics of articulation function disorders indicate degradation of all articulatory functions in patients with ALS. The analysis of the test results showed that the degradation of all articulation functions is not the same. It can be noted that

some of the articulation functions that are components of the process of speech formation deteriorate earlier than others. The study clearly shows that the work of tongue is the first to be degraded. Furthermore, the observed exacerbation of dysarthria was more strongly correlated with increased spasms rather than with weakness, suggesting that more severe forms of dysarthria may be largely due to the slower movement of the tongue's muscle structures before significant loss of muscle strength occurs.

The obtained research results allow the statement that the dynamics of changes in articulation functions which image dysarthria in the course of ALS vary.

The direction of changes in all articulation functions is the same - they all deteriorate. However, the rate of changes is different. The rate of deterioration of the function: language - in all investigated aspects and the intelligibility of spontaneous speech in the group of men, women and between the groups, is similar. Functional degradation of these functions occurs at the earliest and runs in a steady manner.

The rate of changes in other articulation functions: length of phonation, voice, reflexes, respiration, lip movements, jaw, functions of the soft palate, speech intelligibility (words), degrades later.

The above findings, allow taking efforts in order to devise the degradation pattern of particular articulation functions. In my study I have also showed how the course of disorders of articulation functions differs, depending on the sex of a patient.

Within the same study, there are no statistically significant differences in the values of particular articulation functions between women and men. However, the degradation rate of the individual articulation functions for men and women differs. In men, the function of breathing is much earlier in functional decline. Women develop earlier problems with effective swallowing. Research proves that the disorders occur in the weakening of the musculus constrictor palatopharyngeus function. That brings the legitimate reason for the creation of a pattern of degradation of articulation functions in the limb onset of the SLA form, characteristic of a patient's sex.