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Identification of Amyotrophic Lateral Sclerosis Using Functional Scale

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Abstract

Objective: Recent studies widely demonstrate that the pattern of progression in sporadic ALS patients is unsolved mystery, especially what factors influence the progression of the disease, therefore we investigated 60 ALS patients who were evaluated with the ALS Functional Rating Scale-Revised (ALSFRS-R) for disease progression and investigated factors contributing to its rapid rate, directed to The First University Clinic of Tbilisi State Medical University during years 2019-2022.

Methods: Overall 60 patients with ALS were investigated, among them 31 male (51%), 29 female (49%), aged 21-84, Riluzole takers, sporadic ALS, Brain MRI, and electrophysiological studies were done in all patients. Patients were diagnosed with Gold Coast Criteria, and evaluated with the ALS Functional Rating Scale-Revised (ALSFRS-R) Patient survey for factors affecting disease progression was filled with a help of the patient /caregiver (a disease-specific questionnaire was invented for this purpose, investigating probable progression modifiers).

Results: It has been established that recent falls-trauma, surgery, and infection have a drastic impact on disease progression, ALS was found to be a not linear progressive disease and can vary in individuals with ALS, we have found that 5% of patients had improvement in ALSFRS-revised, not explained by medication, nor lifestyle change.

Conclusion: The rate of ALS progression appears to be a mystery by itself, predicting the progression rate, and the factors affecting it would be beneficial for ALS patients and may even bring chances to slow or halt the progression, further research is essential.

Keywords: Amyotrophic Lateral Sclerosis (ALS); ALS Functional Rating Scale-Revised (ALSFRS-R)

Introduction

ALS is a disease of the parts of the nervous system that control voluntary muscle movement. In ALS, motor neurons (nerve cells that control muscle cells) are gradually lost. As these motor neurons are lost, the muscles they control become weak and then nonfunctional, thus leading to muscle weakness, disability, and eventually death. ALS is the most

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common form of motor neuron disease [1-5]. Amyotrophic lateral sclerosis (ASL) is one of the heavy neurodegenerative diseases [2-5]. It constitutes a progressive neuromuscular disease, first records of which have been made in medical literature in the first half of 19th century (Charles Bell-1824; François-Amilcar Aran-1850) [6-9].

The clinical symptoms of the disease, in line with developed neurological deficit, was published by Jean-Martin Charcot in 1869 [3-5,10]. He also was the one who coined the term Amyotrophic lateral sclerosis. This disease was very rare in 19th century and there were only a few methods developed for its diagnostics [3-5,10].

Objective

Recent studies widely demonstrate that the pattern of progression in sporadic ALS patients is unsolved mystery, especially what factors influence the progression of the disease, therefore we investigated 60 ALS patients who were evaluated with the ALS Functional Rating Scale-Revised (ALSFRS-R) for disease progression and investigated factors contributing to its rapid rate, directed to The First University Clinic of Tbilisi State Medical University during years 2019-2022. Methods: Overall 60 patients with ALS were investigated, among them 31 male (51%), 29 female (49%), aged 21-84, Pedigree analysis was also performed: a pedigree was compiled using standard symbols, which includes information on the health status of the proband's relatives. As a result, relatives who are at risk of developing the disease or are carriers of the pathogenic allele have been identified. Such individuals were given appropriate genetic counseling and recommended for genetic testing.

Results

In addition, upon necessity, the risk of developing the disease for the next generation was calculated. 5 (8.33%) patients had confirmed diabetes mellitus, and one (1.6%) patient had a family history of athetosis. 4 (6.66%) patients had to come into contact with poisonous chemicals. 2 (3.33%) patients have a history of brain trauma, 35 patients (58.33%) are tobacco users (Figure 1).

Chart - patients - according to anamnesis:

- 8 (30%) patients have a one-year anamnesis of confirmed ALS disease.
- Three years old - 34 (56.66%) patients.
- 5 (8.33%) patients have a history of 5 years.
- 2 (3.33%) patients have a history of 10 years.
- One (1.66%) patient has an anamnesis of more than 10 years.

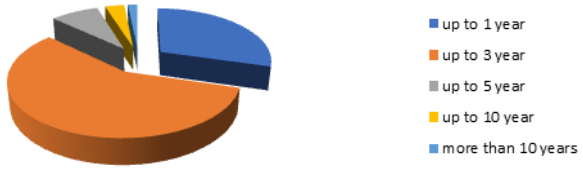

Figure 1: History of patients with confirmed ALS disease.

Patient's functional condition was evaluated by the Amyotropic Lateral Sclerosis Functional Rating Scale (ALSFRS), where the patient's condition is evaluated from zero to four points according to the following symptoms: speech, salivation, swallowing, writing, chewing and processing food (with or without a gastrostomy), dressing and hygiene, turning in bed and dressing, walking, stair-climbing, breathing.

The higher the total score, the more the patient's vital functions are preserved.

According to our research, there are mild functional changes (according to ALSFRS) from 31 points to 40 points in 14 (8.4%) patients, moderate form severity at the time of examination (21 points to 30 points) in 15 (8.4%) patients, severe form (from 10 points to 20 points) with 25 (25%) patients, And 6 (10%) patients with very severe form (from 0 to 10 points), which is depicted on the chart (Figure 2).

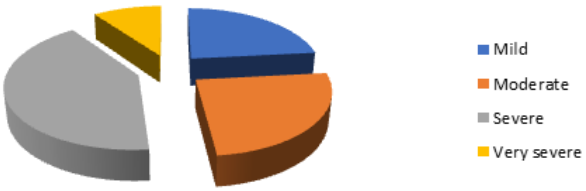

Figure 2: The condition of patients during the examination according to ALSFRS.

The disease started with symptoms - numbness of muscles, weakness of limbs, fasciculations in 36 (60%) patients.

According to our research, the diagnosis of ALS is difficult until muscle atrophy and tremors are detected. ALS disease with 23 (38.33%) patients started with symptoms of one or both legs. Patients felt uncomfortable while walking, the ankle lost its flexibility, its range of motion was limited. Muscle weakness is expressed; muscle spasms; increase of deep reflexes or expansion of the reflexogenic zone; pathological reflexes; pronounced muscle atrophy; increased spasticity; At this time, the upper limbs were less damaged, although the flexibility of the fingers in the upper limbs is limited.

ALS with bulbar events was detected in 24 (40%) patients with difficulty speaking, the patient spoke "through the nose", later had difficulty swallowing. Disturbance of speech (dysarthria, anarthria), voice production disorder (dysphonia, aphonia). Disappearance of soft palate and throat reflexes, salivation, breathing disorders were soon added to the symptoms. In 11 (18.33%) patients with ALS confirmed by us, the symptoms included signs of both lower and upper motoneurons damage, upper motor neurons were expressed: muscle hypertonia, hyperreflexia, Babinski's pathological reflex; and - in case of damage to the lower motoneurons: muscle weakness and atrophy, involuntary fasciculations. According to our study, patients lost their ability to move independently, but their mental abilities were not limited. They survive through artificial lung ventilation and artificial nutrition.

Thus, the frequency and distribution of amyotrophic lateral sclerosis in Georgia from 2016 to date was determined.

Riluzole takers, sporadic ALS, Brain MRI, and electrophysiological studies were done in all patients. Patients were diagnosed with Gold Coast Criteria, and evaluated with the ALS Functional Rating Scale-Revised (ALSFRS-R) Patient survey for factors affecting disease progression was filled with a help of the patient /caregiver (a disease-specific questionnaire was invented for this purpose, investigating probable progression modifiers).

Conclusion

It has been established that recent falls-trauma, surgery, and infection have a drastic impact on disease progression, ALS was found to be a not linear progressive disease and can vary in individuals with ALS, we have found that 5% of patients had improvement in ALSFRS-revised, not explained by medication, nor lifestyle change. The rate of ALS progression appears to be a mystery by itself, predicting the progression rate, and the factors affecting it would be beneficial for ALS patients and may even bring chances to slow or halt the progression, further research is essential.

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