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Clinical and Epidemiological Study of Motor Neuron Disease

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ABSTRACT

Background. According to worldwide data, the average incidence of motor neuron disease (MND) is approximately 0.2–2.4 cases per 100,000 population. However, despite the relative rarity of this disease, its medical and social significance is extremely high: motor neuron disease is an incurable, disabling disease with a high incidence of lesions in people of working age, inevitably leading to death. The disease is based on the combined progressive degeneration of upper and lower motor neurons, which is clinically manifested by a combination of signs of central and peripheral paralysis. The etiology and pathogenesis of the disease remain not fully studied.

Material. In the studied group of patients with MND, the average age was 56.2±9.2 years. The smallest age of the patient was 33 years, the largest was 72 years. Thus, the majority of patients with MND were middle-aged and older, there were only 2 patients under 34 years of age.

Conclusions. Age and sex characteristics, distribution by onset forms, variants and rate of disease progression in patients in the Ferghana population are similar to the data of the European register. Patients with MND lose their ability to work before the final diagnosis is made, their disability is documented with a delay, on average, by 7-8 months. from the moment of actual disability and for 5 months. after the final diagnosis of MND. The results of our analysis of the 2021 GBD may offer objective, recent information for resource allocation and healthcare planning related to MNDs at global and national levels.

Keywords: MND, degeneration of upper and lower motor neurons, amyotrophic lateral sclerosis, muscle lesions, bulbar disorders.

INTRODUCTION

According to 2019 Global burden disease (GBD) estimates, there were ~268,673 [95% uncertainty interval (UI), 213,893–310,663] prevalent cases and 63,700 (95% UI, 57,295–71,343) incident cases of motor neuron disease (MND) worldwide. In 2019, MND caused 1,034,606 (95% UI, 979,910–1,085,401) DALYs and 39,081 (95% UI, 36,566–41,129) deaths worldwide. The global prevalence and deaths due to MND in 2019 were increased (1.91% [95% UI, 0.61–3.42] and 12.39% [95% UI, 5.81–19.27], respectively) compared to 2009, without significant change in incidence.

Amyotrophic lateral sclerosis, progressive muscular atrophy, primary lateral sclerosis, pseudobulbar palsy, spinal muscular atrophy and hereditary spastic paraple-

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gia- were included for analysis as MNDs. Among MNDs, Amyotrophic lateral sclerosis (ALS)—the most common disease entity—causes respiratory failure in 50% of patients within 2 years of diagnosis. Other MNDs also have poor long-term prognoses, imposing a socioeconomic burden on patients and care givers [1, 2]. The basis of motor neuron disease (MND) is the combined progresssive degeneration of the upper and lower motor neurons, which is clinically manifested by a combination of signs of central and peripheral paralysis. The etiology and pathogenesis of the disease remain not fully learnt.

Now, the epidemiological situation of MND in the Fergana Valley is not exactly known, and therefore it seems important to analyze the epidemiological data on MND in Andijan.

FORMS OF MND

In motor neuron disease has several main forms:

• **ALS (Amyotrophic Lateral Sclerosis)** develops in 80% of people with motor neuron disease, causing mainly weakness, cramps and wasting in the muscles, stiffness of the upper and lower extremities;

• **PBP (progressive bulbar palsy)** affects about 10-25% of people, spreads to the neurons of the spinal cord and brain, is manifested by difficulties in chewing, swallowing, slurred speech;

• **Progressive muscle atrophy** (8%) - a rare type of MND, extends to motor neurons in the lower extremities, develops weakness, muscle atrophy, fasciculations and weight loss;

• **PLS (lateral primary sclerosis)** (2%) affects predominantly motor neurons in the upper extremities, developing muscle spasticity, unsteady walking with difficulty speaking.

Both genetic predisposition and environmental factors play a role in the development of MND, and the main risk factors for developing MND are considered to be male sex and age over 50 years [3,7].

It is known that MND in most cases is accompanied by a progressive loss of muscle tissue, which, however, is secondary to degeneration of the anterior horn motor neurons. Whether local muscle necrosis is the primary trigger of the disease remains to be seen.

PURPOSE OF WORK:

To conduct a clinical and epidemiological study of motor neuron disease in the Ferghana Valley based on a personalized register.

METHODS OF RESEARCH

In the studied group of patients with MND, the average age was 56.2±9.2 years. The smallest age of the patient was 33 years, the largest was 72 years. Thus, the majority of patients with MND were middle-aged and older, there were only 2 patients under 34 years of age.

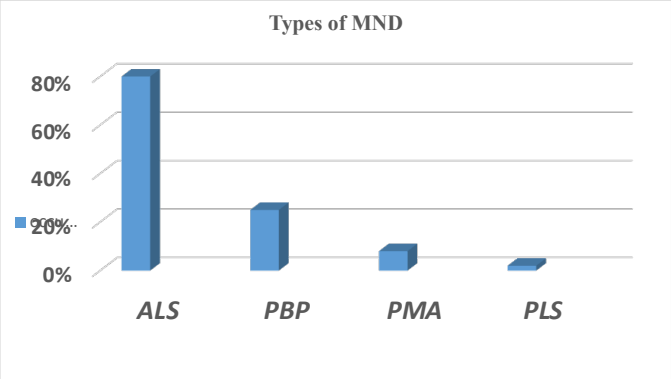

Figure-1. Types of motor neuron disease

RESULTS

Conducted clinical and epidemiological study of MND, and based on the North American classification of ALS [2].

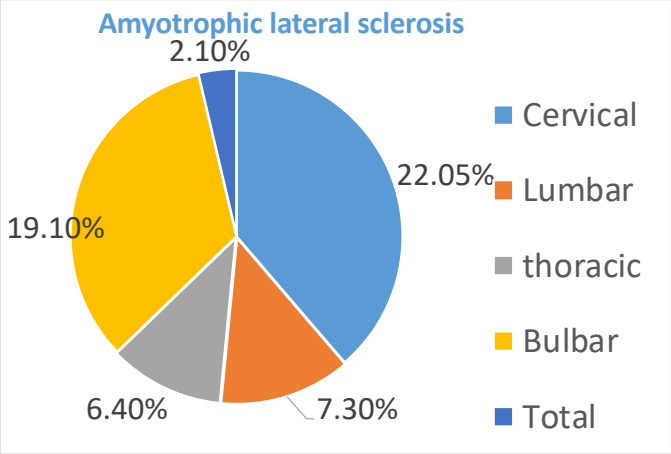

Figure-2. Amyotrophic lateral sclerosis

Identical age-sex characteristics of patients, distribution according to the forms of debuts, variants and rate of progression of the disease in the Ferghana Valley were revealed.

The cervical debut was more often observed - 38%, while the disease began with damage to the muscles innervated by neurons of the cervical enlargement of the spinal cord. With sufficient frequency, the onset of the disease was noted with bulbar disorders (PBP - 32%) and

lumbar lesions (20%) - with primary suffering of the muscles innervated by neurons of the lumbar spinal cord. The diffuse onset of the disease with simultaneous primary paralysis of the muscles innervated by neurons along the entire length of the spinal cord was least often found.

DISCUSSION

Racial diversity and geographic gradients regarding MND incidence were reported in several epidemiology studies [6,8,10]. One study performed in Andijan showed that the risk of ALS was higher in patients than in Namangan and Fergana regions [11]. The age-standardized incidence either decreased or did not change significantly from 2015 to 2021 [4,5]. This phenomenon could be affected by whether accurate or early diagnosis is possible in the area where the incidence is analyzed [8-10].

CONCLUSIONS

Age and sex characteristics, distribution by onset forms, variants and rate of disease progression in patients in the Ferghana population are similar to the data of the European register.

Patients with MND lose their ability to work before the final diagnosis is made, their disability is documented with a delay, on average, by 7-8 months. from the moment of actual disability and for 5 months. after the final diagnosis of MND.

The results of our analysis of the 2021 GBD may offer objective, recent information for resource allocation and healthcare planning related to MNDs at global and national levels.

Ethics approval and consent to participate - All patients gave written informed consent to participate in the study.

Consent for publication - The study is valid, and recognition by the organization is not required. The author agrees to open the publication.

Availability of data and material – Available.

Financing – No financial support has been provided for this work.

Conflict of interest - The authors declare that there is no conflict of interest.

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MOTOR NEYRON KASALLIGINI KLINIK VA EPIDEMIOLOGIK O'RGANISH

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Abstrakt

Butun jahon ma'lumotlarga ko'ra, motor neyron kasalligi (MNK) bilan kasallanish o'rtacha 100 000 aholiga taxminan 0,2-2,4 holatni tashkil qiladi. Biroq, bu kasallikning nisbatan kamdan-kam uchraydiganligiga qaramay, uning tibbiy va ijtimoiy ahamiyati nihoyatda yuqori: motorli neyron kasalligi - davolab bo'lmaydigan, erta nogironlikka olib kelib, mehnatga layoqatli yoshdagi odamlarda ko\p aniqlanib, muqarrar ravishda o'limga olib keladi. Kasallik klinik jihatdan markaziy va periferik falajlik belgilarining kombinatsiyasi bilan namoyon bo'ladigan yuqori va pastki motor neyronlarning birgalikda progressiv degeneratsiyasiga asoslangan. Kasallikning etiologiyasi va patogenezi hanuzgacha to'liq o'rganilmagan.

Kalit so'zlar: MND, yuqori va pastki motor neyronlarining degeneratsiyasi, yon amiotrofik skleroz, mushaklarning shikastlanishi, bulbar buzilishlar.

КЛИНИКО-ЭПИДЕМИОЛОГИЧЕСКОЕ ИССЛЕДОВАНИЕ БОЛЕЗНИ ДВИГАТЕЛЬНЫХ НЕЙРОНОВ

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Abstract

Согласно мировым данным, средняя заболеваемость болезнью двигательных нейронов (БДН) составляет примерно 0,2–2,4 случая на 100 000 населения. Однако, несмотря на относительную редкость этого заболевания, его медико-социальная значимость чрезвычайно высока: болезнь двигательного нейрона — неизлечимое, инвалидизирующее заболевание с высокой частотой поражения лиц трудоспособного возраста, неизбежно приводящее к летальному исходу. В основе заболевания лежит сочетанная прогрессирующая дегенерация верхних и нижних мотонейронов, клинически проявляющаяся сочетанием признаков центрального и периферического параличей. Этиология и патогенез заболевания остаются до конца не изученными.

Ключевые слова: БДН, дегенерация верхних и нижних мотонейронов, боковой амиотрофический склероз, поражение мышц, бульбарные нарушения.