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ПРИОННЫЕ ЗАБОЛЕВАНИЯ

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Прионные заболевания-это неизлечимые нейродегенеративные расстройства, причиной которых является неправильно уложенная, агрегированная последовательность аминокислот, которые называются прионами. Прионные заболевания являются инфекционными и это удалось доказать экспериментальным путем.

Ключевые слова: прион, эпидемия, болезнь, прионные заболевания у людей, история прионных заболеваний, белок.

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PRION SICKNESSES

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Prion diseases are incurable neurodegenerative disorders caused by misfolded, aggregated sequences of amino acids called prions. Prion diseases are infectious and this fact has been proven experimentally

Key words: prion, epidemic, disease, prion diseases in humans, history of prion diseases, protein.

Prion sicknesses

Prion sicknesses - slowly moving sicknesses that impact the human brain. They come from harmful prion proteins. The human brain usually has a prion, it is seen as healthy, as it has a little different shape compared to the harmful one. But when the harmful one gets into the body, it is not killed, and it enters the brain through the blood. There it sticks to neurons and starts to make more of its protein. Because of this, it grows and produces a similar living thing. Also, the harmful prion affects healthy ones, making them change shape and become harmful. Neurons pass away as a result of the harmful prions causing plaques on them.

Most prions are tiny germs. But unlike viruses and bacteria, prions do not share their genes when they spread in and out of hosts. Infection can happen by: blood transfusion, passing from mother to baby, through sex, from food.

Physicians (such as obstetricians-gynecologists, pathologists, and surgeons), veterinarians, laboratory workers, chefs, meat processors, and injecting drug users are at high risk. However, there are situations when prions are inherited or develop spontaneously due to genetic disorders. The progression of the disease depends on the type of prion protein. It takes approximately 10-15 years for symptoms to appear after infection. In simple terms, the disease remains hidden during this time period. This is why it is considered incurable - by the time symptoms emerge, the brain has already suffered extensive damage that cannot be reversed. Encountering someone with prion disease is extremely unlikely as it is a rare condition affecting only 1-15 individuals per million. However, it is important to note that all cases of prion disease result in death within a year of establishing

diagnosis. There have been a few exceptional cases where patients have survived just over 2 years with the illness.

There are 6 known varieties of this pathology today:

1. sporadic Creutzfeldt-Jakob disease (the most common);

2. a new variant of Creutzfeldt-Jakob disease;

3. kuru;

4. Gerstmann-Stroisler-Scheinker syndrome;

5. Alpers disease;

6. fatal familial insomnia.

The first symptoms of these pathologies are: visual impairment; progressive dementia; impaired coordination; speech impairment; anxiety; myoclonia (seizure); depression; tremor; emotional lability (sudden, unbalanced mood swings).

Prion disease causes the following effects:

* Loss of self-criticism and self-care skills

* Increased irritability and demands on the environment, leading to inappropriate behavior

* Memory and cognitive impairment

* Later stages may include dysuria, total muscle rigidity (severe muscle tension that restricts movement), and problems with body areas affected by the disease

* Prion diseases can also result in akinetic mutism (the patient is conscious, hears and understands everything, but cannot speak or move) and eventually lead to coma.

History of Disease Research

The prototype of modern prion diseases is Scrapie disease (Scrapie). Scrapie is a disease of sheep and goats, with symptoms progressing slowly over a period of 2 to 6 years. Specifically, the animals have poor co-ordination of hind limbs, an irresistible urge to rub against objects, and eventually a fatal outcome. Early Cases and Spread one of the earliest recorded cases of scrapie dates back to 1732 in England. In 1755, the disease gained momentum and was so widespread that it became the subject of a petition to the British Parliament by sheep farmers in Lincolnshire. Gradually the disease began to spread in other countries as well, and different names were given to it in different places. Sources report that 34 names are known for scrapie, and only in England the disease has received such names as "fidgety", "rickets", "scratching", "shaking". All the names were intended to convey the animals' desire to rub and scratch themselves. Connection to Other Diseases One of the names of scrapie was "La tremblante", which is very similar in meaning to the disease "kuru". Transmission Experiments In 1899, experiments were carried out and the result was that it was possible to transmit the scrapie to a healthy sheep by infecting it with brain tissue. In 1900, scientists determined that scrapie was a parasitic infection. By 1910, they had solid evidence to

support the transmission of the disease and confirm its infectious nature. This finding was later replicated by researchers in other countries. Initially, studies on scrapie focused on sheep. However, when scientists successfully transmitted the disease to mice, it opened up new opportunities for extensive experimental research. Mice were preferred due to their affordability, abundance, and the ability to measure the disease quantitatively.

Research

The following results were obtained as a result of sociological research carried out among the students of the Bashkir State Medical University , in particular for the sociological survey "What do you know about prion diseases?"

Results analysis:

≈91% of respondents don't know anything about prion diseases.

≈6% of respondents however, have heard it but don't know what it is.

≈3% of respondents are aware that there are some prion diseases.

When asked the question "How did you learn about them?"

90 % said they got their information from internet literature

6 % answered they got their information from reading a book.

4 % wrote their own answers as seen above. Prion disorders, by virtue of their rarity, are not familiar to most students.

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