THE PRION PROTEIN AND ITS EFFECTS ON THE HUMAN BRAIN IN RELATION TO NEURODEGENERATION

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INTRODUCTION

- A prion is an infectious protein responsible for numerous infectious neurodegenerative diseases.
- The word "Prion" was used to distinguish the proteinaceous infectious particles from viruses and viroids.
- The prion was discovered as an accidental byproduct of unsuccessful attempts to unravel the nature of the scrapie agent – for over two decades (Prusiner, 2004).
- The prion was first thought to be a virus, but as further research was made, this idea was revised (Prusiner, 2004).
- The cause of most of the neurodegenerative diseases that are connected to prions are from the protein PrP^C misfolding and becoming the infectious agent known as PrP^Sc (Prusiner, 2004).
- Currently, researchers have settled on the protein only hypothesis (Prusiner, 2004).

OBJECTIVE

- Obtain a better understanding of the prions and to see how prions can affect humans.

METHODS

- Databases for academic journals such as Google Scholar and Ebsco Host were used to gather resources.
- Keywords such as “prion proteins,” “nerves,” & “nervous system” were used to locate sources.
- The book "Prion Biology and Diseases" was also used for reference.

FINDINGS

Various Diseases Associated with Prions

- Scrapie, originally only a disease observed in sheep, was one of the first diseases determined to be caused by prion proteins.
- Various other diseases caused by prions include:
  - Creutzfeldt-Jacob Disease - human-inherited variant of the same condition known as “mad cow disease” (BSE). Contracted by contaminated medical equipment or ingesting contaminated meat. It is often fatal.
  - VPSPr - affects the digestive system
  - GSS - an extremely rare condition that affects the brain
  - Kuru - observed in New Guinea in rare cases; contracted by consuming contaminated human brain tissue
  - Fatal Insomnia - leads to trouble sleeping; it is a hereditary disease but there is a sporadic form that can’t be inherited
  - Chronic Wasting Disease – Rapidly spreading disease affecting Deer, Elk, Moose, and other species (CDC, 2019).

Identification & Treatment

- The most common symptoms that people come in with are confusion, fatigue, hallucinations, and rapid developing dementia. Many of these symptoms are similar to other diseases. MRIs, Spinal taps, blood tests, and neuro exams are used to diagnose prion diseases (John Hopkins, 2019).
- Currently, the timeline for treatment for these diseases is small; found between the initial synapse degeneration and terminal cell loss (Prusiner, 2004).
- In that interval, an effective therapeutic treatment could prevent further degeneration. Once cleared of the infectious proteins, a test of the dendrite trees is done to see if they will regrow (Prusiner, 2004).

CONCLUSION & FUTURE WORK

Over the past few decades, human understanding of the prion protein has increased immensely. They not only have pathogenic, medicinal, and agricultural consequences in the human population, but also have an ecological effect on various species. Medical treatment for these diseases is still advancing as many of the diseases are fatal and have no known cure. Further research should be made into utilizing gene therapy to prevent these diseases on a hereditary basis, as well as more efficient treatment methods of those infected with pathogenic prions.

References


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