PrPST (a Sulfur-based substitute for prP: possible cause of human prion disease pathology [1])

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Published Date: 03-11-2018

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Pathogen latency refers to the period during which a pathogen has the potential to cause disease, and this is largely determined by how quickly the infection subsides. Heterodimeric prion disease is a hereditary PrP pathogen that causes fatal dementia in humans.

Researchers are actively researching how this pathogen is evolving, and the just published paper is an example of such work. Enter PrPST: an insoluble form of prion protein called prPST that appears to protect from a mutant prionâ€™s potent enzyme activity.

PrPST is prion protein deposited in the mouse brain, and over time the prion protein decreases its shear properties in order to form a longer fibre. PrPST is soluble and relatively easy to eliminate from the blood (e.g. conjugated intravenously) as it retains its amino acid complex structure and has a relatively short length. PrPST has been reported previously in reference to prion pathology, but has never been identified as a cause of prion disease. In the present paper, based on observations of prion-associated symptoms in hLD mice, the authors suggest that prPST is an important causal factor in the propagation of a hLD gene variant by preferentially working within this affected gene. The hLD mice develop neurodegeneration, whereas mice in the control group do not.

The authors assert that the protective properties of prPST are more pronounced in animal models of human prion disease, and that the morphological features they describe may, in part, reflect that effect. They state that they conducted their experiments by applying a mutant, hemophiliac, HLD prion model to different mouse species, and also by employing a metabolic pathway restricted to natural prion metabolism.

Due to the lack of robust epidemiological studies, however, the authors caution against recommending that prPST should be used as a treatment in humans. Human clinical trials are currently being undertaken with potentially amenable prion proteins.

The article is of particular interest given the current debate surrounding functional differences between common prion proteins. The authors recommend that further research should proceed into identifying the functional qualities and etiology of prLp, by comparison of the prLp-PI in peripheral blood with the mouse surrogate prLp prP that they have identified.

A Bunch Of Birds That Are Sitting On The Ground