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Exploring the Transmissible Zoonotic Potential of Animal Prion Diseases

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Mad Cow disease is a fatal and serious disease often found in Europe. Mad Cow disease, also known as Bovine Spongiform Encephalopathy, is often characterized by vacuoles found within the cytoplasm of neurons, which often leads to the appearance of a spongy like brain. Some other characteristics of prion diseases are characteristic spongiform changes associated with neuronal loss, long incubation periods, and a failure to induce inflammatory response [1].

There is strong evidence that Bovine Spongiform Encephalopathy is transmitted via a proteinaceous particle known as a prion. Prions are an abnormal, transmissible agent that is able to induce abnormal folding of normal cellular prion proteins in the brain. They are able to self-replicate after an infection by distorting the shape of its native counterpart. Prions initially appear in the tonsils and other lymphoid organs before spreading to the nervous systems. Once it reaches the nervous system, they induce apoptosis of neurons which cause lethargy and erratic behavior in affected cows. Prions diseases are always fatal to the infected host [2]. Prion based diseases such as Mad Cow disease and Creutzfield-Jacob disease are presumed to be transmitted by eating contaminated beef. This was observed for Mad Cow disease where cattle feed that was fed to cows may have contained ruminants of infected sheep, which is thought to have resulted in species crossover of the sheep prion to cattle [3].

Prions are known to cause neurodegenerative diseases in many other species such as deer, sheep, mink and humans. In humans, the disorder is known as Creutzfield-Jacob disease and is relatively rare in occurrence. Many individuals who are infected with the disease are often times over the age of 45, however cases have been found with patients under the age of 25 being infected [4].

Prion diseases lead to devastating and fatal conditions in both humans and animal. Creutzfeldt-Jacob disease as well as the variant form of Creutzfeldt-Jacob disease and Kuru, are among the few diseases found to be caused by prions in humans. The causative agent of prion diseases, which are also known as transmissible spongiform encephalopathies (TSE), is thought to be a prion. However, this is a debated topic as there is research to suggest that TSE are not caused by prions as is generally accepted but is instead caused by viral agents [5]. Dr. Laura Manuelidis is a researcher who is challenging the accepted idea of prions being the sole cause of TSE. In her research, she has stated that there is no reproducible evidence that prion protein can infect animals and humans and likewise does not fulfill Koch's postulates for infection. Her current hypothesis is that host prion is a required receptor for TSE viruses and that viral prion membrane interactions ultimately cause a pathological prion response. In 2006, Dr. Manuelidis published an article which reported that they had found a 25 nm diameter virus like protein in infectious brain fractions with little prion protein. In that paper, she discussed the ideas that in all of her studies and stimulations of the host innate immune response, all pointed towards a foreign pathogen rather then an unpredictable spontaneous mutation within the host prion conformation. She instead discussed how the ability of slow viruses to survive harsh environmental conditions and enzymatic assaults and also to remain hidden in the host and persist for many years all fit nicely with the characteristics of TSE agents [6].

There are also studies testing the notion of other proteins exhibiting prion like transmissions. One such example is the idea that alpha-synuclein behaves like a prion. According to several studies and evidence, alpha-synuclein and prion protein both adopt an alpha helical rich conformation under physiological conditions and are both capable of refolding into a beta sheet conformation. This readily aggregates into oligomers and amyloid fibrils, which are thought to be toxic to the host and capable of causing neurodegeneration as is seen in Parkinson disease. Further research has implied that Parkinson is a prion disorder due to the increased production and impaired clearance of proteins such as alpha-synuclein [7]. This leads to the misfolding and formation of toxic oligomers which aggregate and can lead to cell death.

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