

Researchers embark on work to control the prion epidemic affecting deer in the USA

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Spanish researchers at the Centre for Cooperative Research in Bioscience, CIC bioGUNE, in collaboration with the University of Kentucky (USA), have discovered a new way to control the stability of certain types of prions (the pathogen agents responsible for transmissible spongiform encephalopathies or TSEs) by means of selecting certain proteins. This gives the opportunity to control prion disease in deer, which has become an epidemic in the USA and Canada. The discovery was published last week in the prestigious scientific journal *Science*.

While the most prevalent prion disease in Europe is *scrapie* in sheep and goats, together with the epidemic of [bovine spongiform encephalopathy](#), or [mad cow disease](#), which appeared in the 90s, in the USA, it is the deer prions that are of most concern to the scientific community. Although there is no statistical evidence that deer prions can infect humans, the disease in wild animals is continuously expanding in numerous states in the USA and has already reached some Canadian states.

The chronic wasting disease (CWD) is a transmissible spongiform encephalopathy that affects pet and wild ungulates. Deer prions infect several types of different deer species, the most common being the [mule deer](#) and the Canadian deer. The first signs diagnosed date back to 1967 and were found in a mule deer (*Odocoileus hemionus hemionus*) held in captivity in Colorado (USA).

Epidemiological data suggest that CWD is a self-sustaining disease and it seems that it can be transmitted horizontally in captive populations.

Recent studies indicate that the transmission of prions in wild populations may occur through contaminated urine, faeces and saliva.

Because of the significance of the epidemic, the disease is being researched at the Prion Laboratory at the Proteomics Unit of the CIC bioGUNE, which started its work earlier this year in 2010 at the Bizkaia Technology Park under the direction of Joaquín Castilla, a researcher hired by the IkerBasque Agency, with the main objective of furthering knowledge of transmissible spongiform encephalopathies (TSEs).

The role of prion strains

The findings of the CIC bioGUNE researchers, who were part of an international team coordinated by the University of Kentucky, studied the role of the various properties of the prion strains in the transmission and spreading of a prion disease in depth, something which until now was unknown.

For this study, [transgenic mice](#) susceptible to [prion disease](#) in cervids were designed and generated. For the first time, these animals helped identify two biologically distinguishable strains of CWD prions, which has led to the conclusion that both prion strains are interrelated. Whilst the transgenic mice, designed to replicate Canadian deer prions, showed a stable propagation of each one of the prion strains, the transgenic mice designed to replicate prions from mule deer propagated an unstable mixture of CWD strains, preventing their separation through their biochemical properties.

The significance of this study is due to the fact that it is the first time that a single difference in the primary sequence of amino-acids of the protein responsible for prion diseases (Canadian deer versus mule deer) is shown, allowing the selection of prion strains. This discovery shows new data on the possibility of controlling the stability of some prion

strains by means of modification of the primary structure of the protein responsible. For the first time, the study also shows the existence of at least two types of CWD strains with clearly distinct pathogenic behaviours.

In this last decade much has been achieved in the understanding of prions. In this regard, the epidemic of the bovine spongiform disease in Europe ("mad cow" disease) has sparked an increased in the number of research groups devoted to this type of infectious agents. Nonetheless, nowadays prions are still greatly unknown. According to Castilla, prions are probably "one of the most intriguing pathogenic agents in nature since their alleged composition relating them to only one protein and the emergence of clearly distinct strains give them a scientific value that cannot be compared".

One of the great unknowns in the area of prion diseases and which Prof. Castilla's group devote themselves to is how a single protein is capable of infecting some species and not others. Although great strides in this direction are being made thanks to new techniques that have recently emerged, there is still a long way to go.

Provided by Elhuyar Fundazioa

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