CIC bioGUNE
Biozientzietako Ikerkuntza Kooperatiboko Zentroa
Centro de Investigación Cooperativa en Biociencias

PRESS RELEASE

A new laboratory at CIC bioGUNE will research the mad cow disease

- *The epidemic of bovine spongiform disease in Europe caused the increase of research groups working in this field.*
- *Prion diseases belong to a group of fatal neurodegenerative diseases affecting human beings and animals, for which there is no treatment available so far.*
- *Scrapie and Creutzfeldt-Jacob disease are the two main diseases caused by prions.*
- *A high percentage of all the fatal familial insomnia (a genetic prion disease affecting humans) cases in the world can be found in the Basque Country.*

(*Bilbao, February, 2010*).- The Prion Laboratory of the Proteomics Unit of CIC bioGUNE has started its work on the premises of the Building 502 in the Bizkaia Technology Park. The main objective of this Laboratory is "*to study in depth the transmissible spongiform diseases* (TSEs)", explained Joaquín Castilla, the head of the laboratory.

The main research line will be the study of molecular mechanisms of prion transmission between different species. The researchers of this laboratory want to understand the ways by which prions can infect some species but not others. In order to carry out the research, new molecules that block the replication of prions should be studied and designed, so that they can be used in therapy.

"*Prions are pathogens that cause the TSEs, which are also called prion diseases. TSE diseases belong to a group of fatal neurodegenerative diseases affecting human beings and animals, for which there is no therapy available so far. These diseases can have*

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various origins: hereditary, sporadic (supposedly spontaneous) and infectious", explains Prof. Castilla.

The epidemic of the transmissible bovine spongiform disease in Europe, known as mad cow disease, resulted in the increase of research groups that work on this type of infectious agents. As a consequence, big progress has been made in acquiring more knowledge about prions. However, they are still unknown to a great extent.

In Castilla's opinion, prions are probably "*one of the most intriguing pathogens of nature, since their supposed composition is linked to only one protein and the appearance of clearly differential strains makes them of incomparable scientific value*". Their replication mechanism, which is similar to the development of such diseases as Alzheimer or Parkinson, makes them a unique pathogen. "*Additionally, we still do not know what a prion is, so they are an irresistible subject of study for such people as the researchers of our group and for myself too*", concludes Castilla.

One of the greatest mysteries in the area of prion diseases, on which the group of Prof. Castilla works, is how a single protein can infect some species but cannot infect others. Although great progress is being made in this area thanks to new techniques that have been recently developed, there is still a long way ahead.

One of the techniques that are currently used to study prions is the replication *in vitro*. In the decades of 1980 and 1990 the only techniques used to study the prions and their transmissibility were based on the inoculation of experimental animals with samples that contained the infectious agent. Later, some *in vitro* techniques were created, where the prions were amplified in a test tube. "*Although the bioassays based on genetically modified animals replaced many* in vivo *experiments, nowadays most of the research is based on* in vitro *techniques. We can now generate prions of different species starting from prions of any species. The PMCA technique* (Protein Misfolding Cycling Amplification), *developed by our group, allows us to amplify these infectious agents in the laboratory*", states Castilla.

It is actually an essential technique for the diagnostics as well as for the study of molecular mechanisms through which the prions can replicate.

Scrapie, the disease that affects sheep and goats, is probably the oldest known prion disease. Nonetheless, the BSE (Bovine Spongiform Encephalopathy) is the disease that caught most public attention, due to its implication in the generation of a new disease in humans and its proved transmission to most of other species. There is another disease, the Creutzfeldt-Jacob disease, which causes 1-2 cases per year per million inhabitants and is still unknown to a great extent, especially in its sporadic version, where the origin

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is still a mystery. A high percentage of all the fatal familial insomnia cases in the world, which is a genetic prion disease that affects humans, is located in the Basque Country.