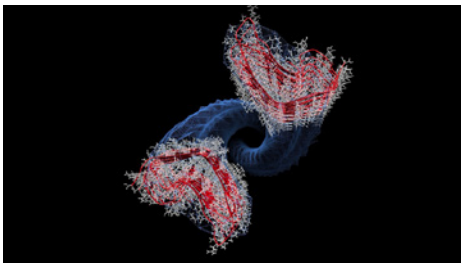

APPLIED RESEARCH

A MODEL EXPLAINS HOW PRION, THE PROTEIN THAT CAUSES MAD-COW DISEASE, REPRODUCES

For the first time, a realistic computational model has been created to explain the reproduction mechanism of prion, a toxic protein that, in the middle of the 1990s, became famous throughout the world as the protein that causes the "mad-cow disease." The study, which has been published in the *journal PLOS Pathogens*, was funded by the Telethon Foundation and conducted by the Istituto Telethon Dulbecco and the University of Trento, in collaboration with the INFN, the University of Santiago de Compostela (Spain), and the University of Alberta (Canada).

Prions are anomalous versions of proteins that are normally found in the brains of mammals, and in other species. They are capable of reproducing and spreading themselves in a similar way to viruses and bacteria. We know that they can induce the change of their normal form to the anomalous one, although the precise mechanism is still unknown. Over time, the anomalous form takes over and forms aggregates that kill nerve cells, provoking extremely serious neurodegenerative conditions called transmissible spongiform encephalopathies. Those we know of that strike humans are Creutzfeldt-Jakob disease, fatal familial insomnia, and Gerstmann-Straussler-Scheinker disease. Bovine spongiform encephalopathy is also very well known, having caused a genuine epidemic among cows beginning halfway through the 1980s, first in England and then in the rest of Europe. Several rare cases of transmission to humans, as a result of ingesting infected meat, have also been recorded.

Researchers have revisited the structure of prions and proposed a new structural model in line with the most up-to-date experimental data. Using a new computational method, derived from mathematical methods that were developed in particle physics, they have, thus, reconstructed the reproduction mechanism. The model will allow researchers to conduct a focused search for drugs that are capable of counteracting serious neurodegenerative diseases that are currently incurable. ■