

New prion protein may offer insight into mad cow disease

Scientists have discovered a new protein that may offer fresh insights into brain function in mad cow disease. “Our team has defined a second prion protein called ‘Shadoo’, that exists in addition to the well-known prion protein called ‘PrP’ ” said Professor David Westaway, director of the Centre for Prions and Protein Folding Diseases at the University of Alberta.

“For decades we believed PrP was a unique nerve protein that folded into an abnormal shape and caused prion disease: end of story. This view is no longer accurate,” Westaway adds.

The study was conducted jointly by the University of Toronto, University of Alberta, Case Western Reserve University (Ohio) and the McLaughlin Research Institute (Montana). The research is published today in the EMBO Journal and represents a culmination of work initiated at the University of Toronto in 1999, and then continued more recently at the University of Alberta.

This is the first discovery since 1985 of a new brain prion protein. “A second prion protein had been inferred by other research, based on indirect studies and the examination of DNA sequences,” said lead author Joel Watts, a graduate student at the University of Toronto’s Centre for Research in Neurodegenerative Diseases. “But we not only demonstrate that this theoretical protein really exists and shares several properties with healthy PrP; we have also defined an unexpected alteration in prion infections.

“As the PrP molecule alters shape and accumulates in a prion-affected brain, the Shadoo protein seems to disappear,” Watts added. Since proteins in a living cell are the molecules “that do the work, this is likely to be significant,” he said.

“Many facets of a prion disease like BSE are puzzling,” Westaway said. “The puzzles include the cause of death of brain cells, the function of normal prion proteins, and the rules governing emergence and spread of prions from animal to animal. We believe the Shadoo protein can give us a fresh purchase on these important questions.”

Source: University of Alberta

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