

New prion protein may offer insight into mad cow disease

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Scientists have discovered a new protein that may offer fresh insights into brain function in mad cow disease. Research led by Dr. David Westaway has led to the first discovery since 1985 of a new brain prion protein.

"Our team has defined a second prion protein called 'Shadoo,' that exists in addition to the well-known prion protein called 'PrP'," said Westaway, a neurology professor and 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 added.

Westaway joined the U of A after leaving the University of Toronto a year ago. The study was conducted jointly by the U of A, U of T, Case Western Reserve University (Ohio) and the McLaughlin Research Institute in Montana. The research is published in the EMBO Journal and represents a culmination of work initiated in 1999.

"It's taken years," Westaway said of the search for a new 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 U of T 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. Because proteins in a living cell are the molecules "that do the work, this is likely to be significant," he said.

Proteins are built up from amino acid building blocks, like beads on a string. However, these strings can fold up into highly complex three-dimensional shapes, called "conformations." Proteins carry out the important activities of living cells. For example, these can serve a structural role as scaffolds, they can act as molecular motors to move other molecules, they can sense the environment near the cell and they can catalyze chemical reactions.

Prions are proteinaceous infectious pathogens that cause diseases like BSE (mad cow disease) and Creutzfeldt-Jakob disease.

"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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