

Prions show their good side

Prions, the infamous agents behind mad cow disease and its human variation, Creutzfeldt-Jakob Disease, also have a helpful side. According to new findings from Gerald Zamponi and colleagues, normally functioning prions prevent neurons from working themselves to death. The findings appear in the May 5th issue of the *Journal of Cell Biology*.

Diseases such as mad cow result when the prion protein adopts an abnormal conformation. This infectious form creates a template that induces normal copies of the protein to misfold as well. Scientists have long assumed that prions must also have a beneficial side but have been unable to pinpoint any such favorable traits.

In the new work, the authors found that mice lacking the prion protein had overactive brain cells. Their neurons responded longer and more vigorously to electrical or drug-induced stimulation than did neurons that had normal prion protein. This hyperactivity eventually led to the neurons' death. The results might help explain why misfolded prions cause dementia: in the wrong conformation, the prion can no longer protect brain cells from deadly overexcitement.

Source: Rockefeller University

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