Sequence and structure key to prion disease transmission

June 14 2010

Prion diseases are lethal neurodegenerative disorders that include Creutzfeldt-Jakob disease (CJD) in humans and bovine spongiform encephalopathy (BSE; commonly known as mad cow disease) in cows. A team of researchers, led by Adriano Aguzzi and Christina Sigurdson, at Universitäts Spital Zürich, Switzerland, has generated data in mice that provides greater understanding of the factors that determine how easy it is for prion diseases to be transmitted to a new host species.

This information provides new insight into a highly important food safety issue; dietary exposure to beef contaminated with the BSE agent is believed to have caused nearly 200 cases of variant CJD in humans.

The key infectious agent in prion diseases is PrPSc, a highly aggregated form of the cellular prion protein (PrPC). The ease with which prions from different species can be transmitted to a new host species varies dramatically.

The team found that transmission between species with the same protein building block at position 170 in PrPC was relatively easy while it was relatively difficult between those species with different building blocks at that position.

As this protein building block influences the structure of the PrPC protein, the authors suggest that local structure of PrPC affected by the protein building block at position 170 might have a triggering role in prion transmissibility between different species.

More information: View this article at:
www.jci.org/articles/view/4205 … 456180f4a34aad821c6f

Provided by Journal of Clinical Investigation