

# Is there more to prion protein than mad cow disease?

September 30 2008

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Prion protein, a form of protein that triggers BSE, is associated with other brain diseases in cattle, raising the possibility of a significant increase in the range of prion disease. Publishing their findings in the open access journal *BMC Veterinary Research*, scientists have detected changes in the production and accumulation of the prion protein in the brains of cattle with a rare neurodegenerative disorder.

Martin Jeffrey of the Veterinary Laboratories Agency led a research team that tested 15 brains of cattle with idiopathic brainstem neuronal chromatolysis and hippocampal sclerosis (IBNC). They are the first group to show that the brains of animals with this disease accumulate prion protein (PrP), the protein that misfolds to cause BSE and which, when transmitted to humans through the food chain, can cause the deadly Variant Creutzfeldt-Jakob disease

IBNC is a rare neurological disease of adult cattle. It was first characterised in 1988 following laboratory tests from cattle suspected of having BSE. Although IBNC has some clinical similarities to BSE, the brains of affected cattle do not have the neuronal vacuolation (lesions) typical of BSE.

Further laboratory tests suggest that the misfolded form of PrP, which accumulates in the brains of BSE cases, is not present in IBNC cases. Commercial BSE testing kits did not detect the telltale, BSE-inducing form of PrP either. However, the presence of increased levels of PrP was detected.

"We've shown for the first time that prion protein is somehow involved in IBNC," says Jeffrey, "In this disease, there is an association with abnormally high levels of a prion protein in the brain but clearly this PrP is in a different form to that involved in BSE and CJD. This may have implications for diagnosis and recognition of typical forms of BSE as well as the related diseases in sheep, deer and in man.

Source: BioMed Central

Citation: Is there more to prion protein than mad cow disease? (2008, September 30) retrieved 9 April 2024 from <https://medicalxpress.com/news/2008-09-prion-protein-mad-cow-disease.html>

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