

Advances in knowledge about prions will potentially change theories on prevention and control of diseases like BSE

February's Prion Disease Update on *ProMED* 2012(02) provides an insight into advances in understanding the world of prion diseases. The new knowledge, if substantiated, will certainly direct future efforts to prevent and control a range of neurodegenerative diseases associated with abnormally folded protein.

Prion diseases such as bovine spongiform encephalopathy (BSE) and variant Creutzfeldt-Jakob disease (vCJD) are able to jump species much more easily than previously thought. A study published in *Science* today [26 Jan 2012] shows that in mice, prions introduced from other species can replicate in the spleen without necessarily affecting the brain. The study reinforces the concern that thousands of people in the United Kingdom might be silent carriers of prion infection, potentially able to pass a lethal form of the disease to others through surgery or blood transfusions.

A blood test for variant CJD is for the 1st time being offered to patients from around the United Kingdom and some from abroad who are suspected of having what was once known as mad cow disease. Media in the UK learned that a notification has been sent to neurologists around the country from the NHS [National Health Service] National Prion Clinic and the Medical Research Council's Prion Unit saying that the blood test is now available. Between 5 and 10 samples a week are now being sent in from here and other countries where there have been cases of vCJD. A crucial new phase in assessing the blood test involves 5000 anonymous samples from the American Red Cross that will enable scientists to examine false positive rates.

Scientists from the Florida campus of The Scripps Research Institute have identified a single prion protein that causes neuronal death similar to that seen in bovine spongiform encephalopathy (BSE), but is at least 10 times more lethal than larger prion species. This toxic single molecule or "monomer" challenges the prevailing concept that neuronal damage is linked to the toxicity of prion protein aggregates called "oligomers." In addition to the insights it offers into prion diseases such as BSE and variant Creutzfeldt-Jakob disease, the study opens the possibility that similar neurotoxic proteins might be involved in neurodegenerative disorders like Alzheimer's and Parkinson diseases.