Diagnostics for Prion Diseases

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Background
Detection of the pathological form of the prion protein had been developed in the early eighties, but the technology was never used for screening larger numbers of animals. In 1996, we developed at the University of Zürich with the financial aid of the Swiss National Science foundation and, in particular, the SPP Biotech a first routinely applicable BSE test system. Out of this research, the company Prionics was founded which now pursues the further development and sale of diagnostic products in the field of prion diseases, e.g. Creutzfeldt-Jakob (CJD) in humans, Bovine Spongiform Encephalopathy (BSE) in cattle, and scrapie in sheep.

The Prionics BSE/Scrapie Test
The Prionics BSE/Scrapie screening-test (named Prionics Check) is a rapid diagnostic system for the detection of prions. The test system works directly with homogenized CNS tissue, no further purification or concentration steps are necessary. The detection of prions is based on the immunological detection of the PrP27-30 molecule which represents the protease resistant part of the infectious particle. PrP27-30 is identified in an optimized Western blotting procedure by two criteria: 1. by the binding of the proprietary antibody 6H4 and 2. by the characteristic size of PrP27-30 (27-30kD). The Prionics BSE/Scrapie-Test can be performed directly with tissue homogenates in only a few hours, combining the reliability of Western blotting with the speed needed for mass screening. These advantages are primarily the result of (a) the unique properties of the homogenization and protease incubation buffers and (b) the high affinity and specificity of the monoclonal antibody used.

Active Surveillance
Surveillance represents a critical step to determine the frequency and spreading of prion diseases. Scrapie in sheep has been known for more than 250 years and was always regarded as an animal disease without inpact onto the human population. A number of epidemiologic studies did not find any correlation of the occurrence of scrapie in sheep and its human counterpart, Creutzfeldt-Jakob disease (CJD). This changed upon the discovery of the new variant of CJD (vCJD) which subsequently was linked to transmission of BSE to humans. Transmission of BSE across the species barrier therefore started the need to test for BSE for consumer safety i.e. to prevent as much as possible the spreading of prions to humans.

Surveillance initially meant the observation of clinical signs and subsequent reporting (passive surveillance) which obviously was limited in its efficiency through a number of economic and social pressures. The introduction of active surveillance (i.e. random testing of animals) using The Prionics BSE test by the Swiss Veterinary Authorities clearly showed that a number of BSE animals ended up in the food chain.

Widening the surveillance to the whole of Europe now has led to the discovery of BSE in almost every European country (with the exception of Sweden and Norway) as well as in Eastern European countries and Japan. Currently most animals over 30 months (in some countries over 18-24 months) are being tested for BSE before entering the food chain.