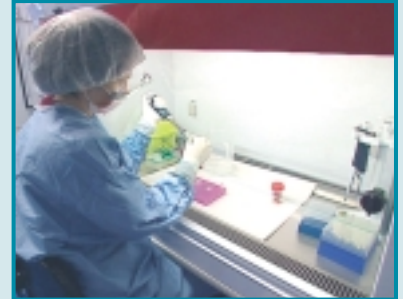


# The diagnosis of prion diseases

Initially perceived as a typically British problem, mad-cow disease became a European problem, in 1996, when the transmissibility to human was demonstrated. The crisis was increased in November 2000, when it clearly appeared that the whole European flock was concerned. Since the beginning of the epidemic, the European Commission was in charge of the dossier implementing the various measures aimed to limit the spreading of the disease or to protect European consumers. This includes the ban of meat and bone meals, massive culling and the removal of specified risk materials.

Since 1999, new tests allowing the post-mortem diagnosis of BSE were introduced. Initially used in the framework of epidemiological studies, they are now systematically used on cattle over 24 or 30 months entering into the food chain. In 2001, 8.5 millions of tests were performed allowing the identification of more than 1000 infected animals.

CEA played a critical role in the fight against BSE by developing a rapid test which is now marketed by the Bio-Rad company under a CEA licence. **This test which is known as the more sensitive is also the more widely used in the world (more than 60 % of the world-wide market).** Since 2002, it is also currently used for the post-mortem diagnosis of scrapie in sheep and goat.



## ■ Towards an ante-mortem test

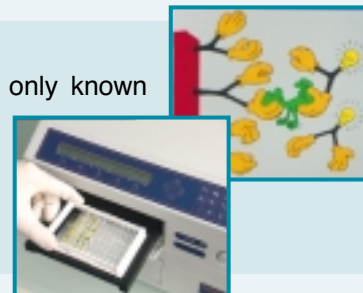
CEA teams remain actively involved in this field of prion disease diagnosis by working on the development of test for preclinical and ante-mortem diagnosis of the variant of the Creutzfeldt-Jakob disease.

## ■ A post-mortem diagnosis of BSE

**Principle:** detection of the abnormal protease-resistant form of the prion protein (PrPres, the molecule marker for BSE) after elimination of the normal form by a proteolytic treatment followed by an immunometric assay.

**First step:** purification of the protease-resistant prion protein (PrPres), 30 minutes.

**Second step:** detection of PrPres by a two-site immunometric assay using monoclonal antibodies, 2h30.



## ■ Rapid test for the preclinical postmortem diagnosis of BSE

- Evaluated by European Commission (DG 24, world invitation to tenders, June 1999) among the four tests evaluated this test was found to detect the marker of cow disease at the lowest concentration and with a great specificity.

- The sensitivity of this test is comparable to the limit of detection of infectivity in the conventional mouse bioassay (Nature, 25 January 2001).

- This test has the capacity to detect PrPres at least three months before the onset of clinical symptoms (Veterinary Record, November 2001).

