Vet. Res. (2008) 39:51

DOI: 10.1051/vetres:2008028

Foreword

It is now approximately twenty years since the first reports in the UK of Bovine Spongiform Encephalopathy (BSE) brought the previously obscure 'prion diseases' into the world spotlight. Initially considered purely an animal health problem, BSE was reported in 1996 to be a zoonotic agent giving rise to variant Creutzfeldt-Jakob disease (vCJD) in humans. To date, vCJD has been responsible for the deaths of about 200 people in Europe.

In the period between the first report of BSE in 1986 and the generalized implementation of measures for preventing human exposure, approximately 400 000 incubating animals entered the food chain. Despite such massive exposure conditions the epidemic in Human and other animal species, predicted by some observers, has not occurred; this situation may have been tragically different but for the existence of the species barrier which hampers transmission of prions between species.

Following the emergence of vCJD and BSE identification in most EU countries, an unprecedented crisis in consumer confidence developed – considerably amplified by media. As a consequence, not only was the ruminant breeding industry brought to near collapse but the very foundations of the EU food protection system were threatened. As a positive collateral effect of the crisis, massive funding was made available for prion research. Thus from just a handful of workers, the prion field became the area of investigation for hundreds of scientists. Their efforts resulted in more progress in TSE comprehension over the past two decades than in the whole of the previous century.

This journal issue, rather than attempting to present a full inventory of prion knowledge, aims to focus on some of the major scientific problems of the field. It provides an up to date vision of

- the current prion fundamentals, from the nature of the infectious agent, to the species barrier phenomenon and the mechanisms that might explain the final neurodegenerative process;
- the extraordinary progress of the methodology – and also the limits – that have been made available over the last decade to investigators of the TSE agents;
- the rational, success and limits of genetic selection as a tool for controlling and eradicating TSE in animals populations;
- and finally the current situation and future perspective of BSE epidemics and of emerging TSE agents such as CWD and atypical scrapie .

Today, classical BSE is under control and TSE seems of no more of interest to both public opinion and policymakers. Resolving the numerous mysteries of prions will rely on those scientists who are able to continue their investigations after the withdrawal of the funding tidal wave over the last decades.

We hope this special issue will assist the reader in putting into perspective the current opinions and facts within the area and highlight the major scientific and public health issues that prion that still require elucidation.

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