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## **BSE/TSE- update of scientific facts**

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Transmissible spongiform encephalopathies (TSEs) or prion diseases include Creutzfeldt-Jakob disease (CJD) in sporadic, accidentally transmitted (iatrogenic), and familial/genetic forms in humans, and scrapie, chronic wasting disease, and BSE and its derivatives in animals. The catastrophic epidemic of BSE in the UK has spilled over to many European countries, Israel and Japan. In spite of highly publicized opinions on the contrary, variant CJD (vCJD) is virtually certain to derive from BSE, but still uncertain in terms of the size of a possible epidemic. As infectivity in vCJD is much more widespread than in other human TSEs, current concerns about possible secondary transmissions, e.g. by blood and blood products, need to be addressed. Definite diagnosis in TSEs still relies on neuropathology or immunochemistry for the prion protein (PrP) performed on tissue from dead individuals, although important inroads into *in vivo* diagnostic testing on body fluids have been recently made. Human disease features a wide and steadily growing spectrum of clinical and pathological phenotypes and PrP gene (*PRNP*) genotypes. Molecular factors determining the considerable phenotypic variation of human TSEs include the *PRNP* genotype, in particular at the polymorphic codon 129, and the “fingerprint” pattern of disease-associated PrP on Western blots from affected brains. However, the pathogenic action of these and other molecular factors has remained unclear, as does the nature of the infectious agent (prion).