SUPPORTING INFORMATION & RISK ASSESSMENTS

TABLE 1. Important TSEs and their characteristics

TSE (host)	Clinical features	Usual transmission	Mean incubation period	Experimental transmission to
Scrapie (sheep)	Behavioural disorder & ataxia	"Spontaneous" – possibly by milk	Usually more than 6 months	Goats, mice and other species
BSE	Behavioural disorder & ataxia	Bovine meat and bone meal to calves	5 years (range 2-10)	Mice, sheep, goats
CWD[67] (deer & elk)	Behavioural disorder & ataxia	"Spontaneous"	Several years	Ferrets, monkeys, goats
HUMAN				
Kuru	Ataxia & terminal dementia in Fore	Oral (cannibalism)	10-12 yrs (range 4-40)	Primates & others
vCJD ('human BSE")	Behavioural disorder & dementia in younger persons	Oral (BSE contaminated food)	16-17 years (range 4-30?)	Humanised mice and other species[68]
Sporadic CJD	Dementia in an older person	"Spontaneous"	?	Primates and other species
Iatrogenic CJD	Progressive dementia (usually in adults)	Grafts of cornea or dura mater or hormones or instruments contaminated with CJD material	Range 1.5 - 30 years	Primates and other species
Familial CJD	Progressive dementia	Associated with inherited mutations in PrP genes[69]	?Lifetime	Primates and other species

[68] Inadvertent secondary transmission to humans by blood transfusion, with a mean incubation period of perhaps 7-8 years.

[69] Other mutations in PrP genes cause other familial neurological disorders such as GSS or FFI.

Fig. 1. BSE surveillance in UK and rest of EU by year

Chart B4: Evolution of BSE cases detected by passive surveillance and active monitoring in the UK

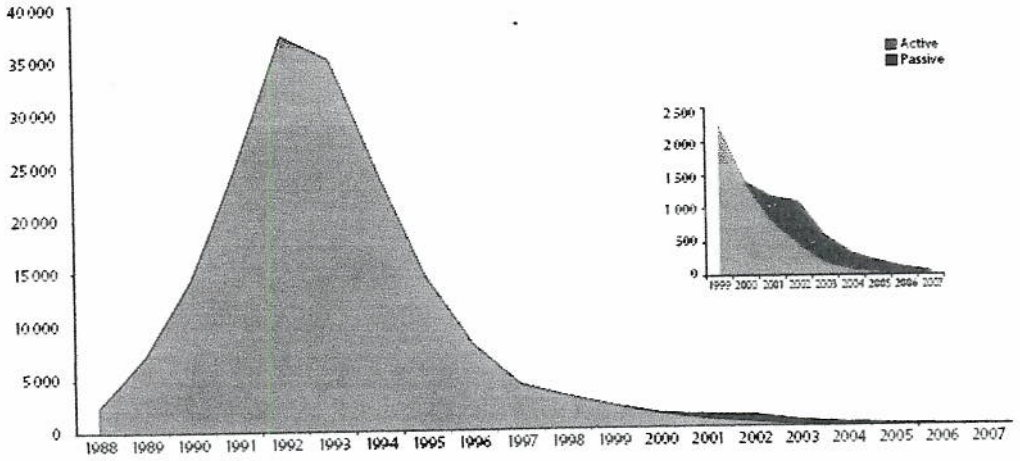
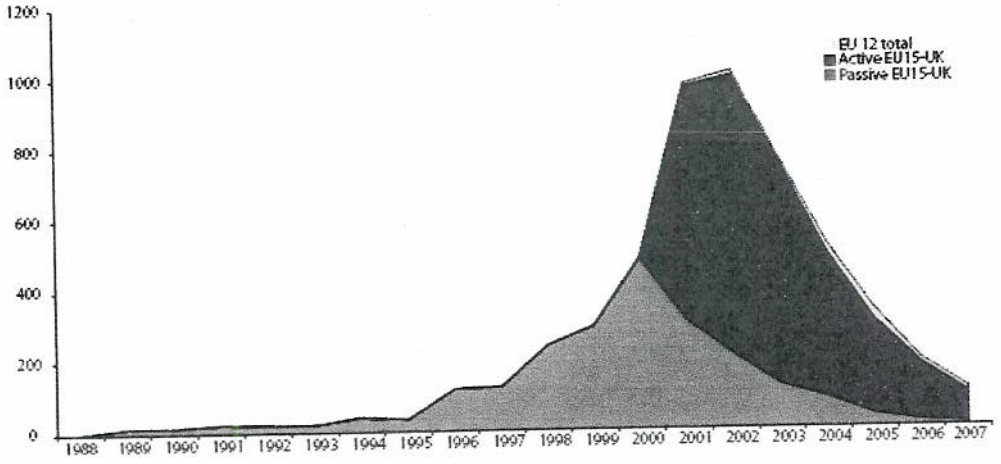
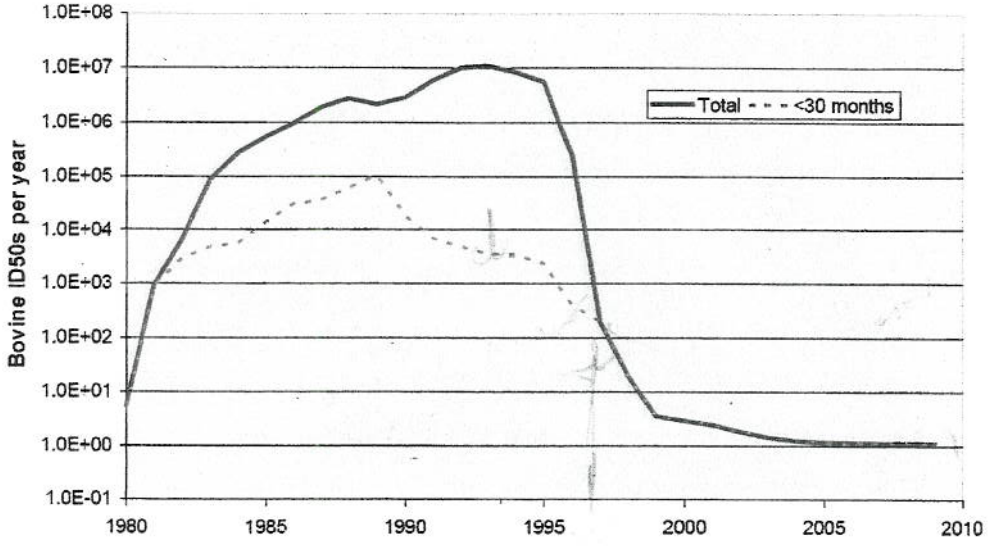


Chart B5: Evolution of BSE cases detected by passive surveillance and active monitoring in the rest of the EU



From the *"Report on the monitoring and testing of ruminants for the presence of TSE in the EU in 2007"*. See http://ec.europa.eu

Fig. 3. Bovine Infective Units (ID50) Entering the UK Food Chain by Year



Note the logarithmic scale of doses, so that the amount of infective material entering the food chain in 1993 is about 10 million times greater than in 2006. The dotted red line shows the contribution from younger animals (less than 30 months at slaughter), and the gap between that line and the top blue line shows the contribution from older animals. As older animals were removed from the food chain by 1997, the lines converge in later years. From Comer and Huntly (2003).

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Transmission of scrapie and sheep-passaged bovine spongiform encephalopathy prions to transgenic mice expressing elk prion protein.

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Chronic wasting disease (CWD) is a transmissible, fatal prion disease of cervids and is largely confined to North America. The origin of CWD continues to pose a conundrum: does the disease arise spontaneously or result from some other naturally occurring reservoir? To address whether prions from sheep might be able to cause disease in cervids, we inoculated mice expressing the elk prion protein (PrP) transgene [Tg(ElkPrP) mice] with two scrapie prion isolates. The SSBP/1 scrapie isolate transmitted disease to Tg(ElkPrP) mice with a median incubation time of 270 days, but a second isolate failed to produce neurological dysfunction in these mice. Although prions from cattle with bovine spongiform encephalopathy (BSE) did not transmit to the Tg(ElkPrP) mice, they did transmit after being passaged through sheep. In Tg (ElkPrP) mice, the sheep-passaged BSE prions exhibited an incubation time of approximately 300 days. SSBP/1 prions produced abundant deposits of the disease-causing PrP isoform, denoted PrP(Sc), in the cerebellum and pons of Tg (ElkPrP) mice, whereas PrP(Sc) accumulation in Tg mice inoculated with sheep-passaged BSE prions was confined to the deep cerebellar nuclei, habenula and the brainstem. The susceptibility of 'cervidized' mice to 'ovinized' prions raises the question about why CWD has not been reported in other parts of the world where cervids and scrapie-infected sheep coexist.

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Prion Infectivity in Fat of Deer with Chronic Wasting Disease

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Chronic wasting disease (CWD) is a neurodegenerative prion disease of cervids. Some animal prion diseases, such as bovine spongiform encephalopathy, can infect humans; however, human susceptibility to CWD is unknown. In ruminants, prion infectivity is found in central nervous system and lymphoid tissues, with smaller amounts in intestine and muscle. In mice, prion infectivity was recently detected in fat. Since ruminant fat is consumed by humans and fed to animals, we determined infectivity titers in fat from two CWD-infected deer. Deer fat devoid of muscle contained low levels of CWD infectivity and might be a risk factor for prion infection of other species.

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