Human prion diseases: epidemiology and integrated risk assessment

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Human prion diseases are devastating and incurable, but are very rare. Fears that the bovine spongiform encephalopathy epizootic would lead to a large epidemic of its presumed human counterpart, variant Creutzfeldt-Jakob disease (vCJD), have not been realised. Yet a feeling of uncertainty prevails in the general public and in the biomedical world. The lack of data on the prevalence of asymptomatic carriers of vCJD compounds this uncertainty. In addition to this problem, Switzerland is currently faced with another issue of major public concern: a recent rise in the incidence of CJD. Here we examine the plausibility of several scenarios that may account for the increase in CJD incidence, including ascertainment bias due to improved reporting of CJD, iatrogenic transmission, and transmission of a prion zoonosis. In addition, we present the design and current status of a Swiss population-wide study of subclinical vCJD prevalence.

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