



INCIDENCE OF CREUTZFELDT-JAKOB DISEASE IN SWITZERLAND.

Issue

1. At the November 2003 meeting the committee requested an update on the experimental research and epidemiology of sporadic CJD (sCJD) in Switzerland. Dr Markus Glatzel from the Institute of Neuropathology and National Reference Center for Prion diseases, Zurich and Dr Lorenz Amsler an epidemiologist from the Swiss Federal Office of Public Health (SFOPH), in the Division of Epidemiology and Infectious Diseases, Bern, have kindly agreed to update the committee on their work.
2. Glatzel *et al* (2002)¹([Annex 1](#)), published in the Lancet, a research letter describing an investigation of the increased incidence of sCJD observed in Switzerland in 2001. Glatzel *et al* (2003)² ([Annex 2](#)) is a review article, published in the Lancet (Neurology) which speculates for the increase in cases and work in progress.

Background

3. The worldwide incidence of sCJD is reported as an estimate of one in a million cases per year. Glatzel *et al* (2002) report that between 2001 and (up to) March 2002, 26 sCJD cases (male n=15 and female n=11) were diagnosed in Switzerland, which represented a yearly incidence of 2.6 deaths per million. There were 19 cases each in 2001 and 2002. The mean age at death was 67.2 years (range 48-83) and the median disease duration was 3.1 months (range 1.3-11.0). The range figures indicate the low and highest values around the mean or median. Unpublished work shows that in 2003, there have been 15 cases of sCJD cases in Switzerland, which represents an incidence of 2.1 deaths per million.
4. All of the variant CJD cases in the UK which have been genotyped have shown to have methionine homozygosity; that is they possess coding for the amino

¹ Glatzel M, Rogivue C, Ghani A, Streffer JR, Amsler L, Aguzzi A. Incidence of Creutzfeldt-Jakob disease in Switzerland. Lancet. 2002 Jul 13;360(9327):139-41.

² Glatzel M, Ott PM, Linder T, Gebbers JO, Gmur A, Wust W, Huber G, Moch H, Podvinec M, Stamm B, Aguzzi A. Human prion diseases: epidemiology and integrated risk assessment. Lancet Neurol. 2003 Dec;2(12):757-63.

acid, methionine on each copy of the prion gene. Of the 25 sCJD cases tested in Switzerland (out of a total of 26), methionine homozygosity was the most common genotype (n=19), followed by valine homozygotes (n=5) and heterozygotes (n=1). Prion proteins may also be classified according to the number of attached carbohydrate groups (glycotyping). Glycotyping of the sCJD patients revealed that type 1 and 2 were the most common (n=17). Nine patients were type 3. None of the cases had a type 4 glycotype profile for PrP^{sc} characteristic of vCJD which is concurrent with the clinical diagnosis that these patients had sCJD.

5. Glatzel *et al* (2003) is a review article that includes data from the Lancet paper of 2002. The authors (Glatzel *et al* 2002 & 2003) provide five possible explanations for the observed increase incidence of sCJD cases in 2001 which has been maintained in 2002. These include: (1) a statistical fluctuation, (2) ascertainment bias after CJD became a notifiable disease in 1999, (3) genetically determined clusters of familial CJD, (4) iatrogenic transmission, or (5) exposure to a TSE agent via the food chain, such as BSE.
6. Since surveillance started in 1989, there have been 451 cases of BSE in Switzerland³. The greatest number (n=68) was reported in 1995. As of 30th September 2003⁴ there have been no cases of vCJD in Switzerland.
7. The SFOPH has asked the national reference centre for prion diseases, (NRPE) to conduct a cross-sectional, 'linked anonymised' prevalence study of PrP^{sc} infection in Switzerland. This will involve the collection of lymphoid tissue from tonsillectomies and autopsies from 15,000 patients. The Swiss authorities considered that the development made in the therapy of prion diseases might allow for treatment if a high prevalence of subclinical infection is found in the Swiss population.
8. An unlinked, anonymised prospective study of tonsillectomy samples for PrP^{sc} is ongoing in the United Kingdom. Dr John Stephenson from the Department of Health will provide the committee with an update on the UK prospective study.

Advice sought from the Committee

The committee is asked to consider the following questions.

1. Can members comment on the proposed five hypotheses for the increased incidence in sCJD?
2. Does the surveillance work in Switzerland have any implications for the UK surveillance programme?
3. Does the committee wish to recommend any further research?

³ [Office International des Epizooties \(OIE\)](http://www.oie.int/)

⁴ <http://www.eurocjd.ed.ac.uk/results.htm>



**Glatzel M, Rogivue C, Ghani A, Streffer JR, Amsler L and Aguzzi A.
Incidence of Creutzfeldt-Jakob disease in Switzerland. *Lancet*. 2002;
360(9327): 139-41.**

**RESERVED BUSINESS
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ANNEX 2**



Glatzel M, Ott PM, Linder T, Gebbers JO, Gmür A, Wüst W, Huber G, Moch H, Podvinec M, Stamm B and Aguzzi A. Human prion diseases: epidemiology and integrated risk assessment. *Lancet Neurol.* 2003; 2(12): 757-63.