

Comment

BSE/vCJD: A calm in the storm?

● Please note that views expressed in *Comment* do not necessarily reflect official policy of the SGM Council.

Sources of data

Worldwide BSE cases:
<http://ourworld-top.cs.com/j1braakman/BSE.htm>

UK cases of CJD month by month
<http://www.doh.gov.uk/cjd/stats/jan01.htm>

Treatments for TSEs on the way:
<http://sparc.airtime.co.uk/bse/Treatmentall.html>

Diagnostic test systems being developed for BSE/vCJD:
www.airtime.co.uk/bse/adco.htm

Further reading

Doh-Ura, K., Iwaki, T. & Caughey, B. (2000).

Lysosomotropic agents and cysteine protease inhibitors inhibit scrapie-associated prion protein accumulation. *J Virol* **74**, 4894–4897

Rabenau, H.F., Cinatl, J. & Doerr, H.W. (eds). *Prions: A Challenge for Science, Medicine and Public Health Systems*. Basel: Karger.

Dealler, S.F. (2001). Should young UK cattle be considered free of BSE or is it endemic? *Br Food J* **4**, 264–280.

While BSE fades, the vCJD is rising. I will try to explain how good news may in fact be round the corner.

Clinical cases of bovine spongiform encephalopathy (BSE) first certainly appeared in small numbers in the UK in around 1983 although we did not realise it until 1986, and even then it was thought an interesting but rare condition. It was only when we could see the numbers rising rapidly that we understood just how serious it was. We now know that over a million cattle were infected, that we ate over 80% of them and that only around 181,000 cattle have been seen as clinical cases. On average everyone in the UK has eaten 50 meals made of the tissues of infected cattle. The number of infected humans could be somewhere between 1,000 and ten million. The first variant Creutzfeldt-Jakob disease (vCJD) cases (now known to be infected with the same strain of prion as BSE) appeared in late 1995. The incidence increased dramatically in 2000 and again in 2001, as if entering an epidemic rise. Predictions that human cases would peak in the next 3-5 years might indicate the total number to be low but other calculations suggest a much later peak will be reached between 2010 and 2020, followed by a long slow decline, resulting in a higher number of cases.

We are now at the point in the UK where BSE cases have faded to around 50 per month, with few animals born (so far) after the 1996 ban on bovine materials ending up in cattle food. Congratulations and a lot of crossed fingers in the meat industry tell us that BSE in the UK is on its way out. Look across the Channel, however and the numbers are rising rapidly. In Germany they only really stopped feeding infectious material to cattle in October 2000 so their epidemic peak will not be reached until 2005. BSE cases have been incubating in large numbers in Europe for the last 5 years and meal made from unwanted bovine products to feed cattle has been exported around the world. BSE is now appearing further and further away with cases reported in Spain, the Czech Republic, Greece, and Hungary. The Food and Agriculture Organization has recently issued an international message telling everyone to stop feeding cattle with bovine products, no matter

what the source, as it expects BSE to be present everywhere in small amounts already.

So at the moment there is calm here in the UK but a storm is brewing: vCJD cases are rising but the cases currently without symptoms are the problem. In such people we must assume infectivity to be throughout the body. For example, vCJD prions may be in their blood and it follows that current UK blood donations may be from an infected person; also the prions will be in body tissues and so surgical instruments will be contaminated after an operation on an infected person. The cost of discarding surgical equipment and importing blood products is currently being covered by Government funding. What is needed desperately is a diagnostic test but if we had one, potentially large numbers of people would be told they are to die of vCJD with no treatment available.

Well, all that was until very recently! Prusiner's group in California demonstrated an old anti-malarial, quinacrine, to be active against the build up of prions in cell cultures. A Warrington 20 year old, Rachel Forber, diagnosed with vCJD, and told that there was no treatment, went over to San Francisco and was treated aggressively with the drug. Her father is determined that she is improving. A second patient started the drug in London at the end of August and full-scale clinical trials are being organized in the UK.

The diagnostic field for BSE/vCJD is not looking quite so blank as it had been only 3 months ago either. One group in Geneva has managed to get prions to multiply in test tubes, potentially to numbers large enough for our current tests to find them. The inventor tells the media that he is hoping the process will be available in a few years. In the meantime, Ruth Gabizon's group in Israel has shown an altered form of the prion to be present in the urine early in the incubation period of mice. She is expecting the test to be available within a year. In London a company is now predicting its test of blood to be available around the same time and that it will be adequately practical for the screening of large numbers of samples.

Is there light at the end of the tunnel? Will we test all the cattle we eat, and screen people before surgery or dentistry? Will we be able to avoid all the CJD risks from blood transfusion and treat the asymptomatic blood donors that we find with the tests? My own opinion is: Yes, the good news in on the way. We may end up testing everyone, but at the moment the UK owes the world a lot for our BSE so we must not stop now.

● **Dr Stephen Dealler is Consultant Medical Microbiologist at Burnley General Hospital, Burnley BB10 2PQ. He has been working on BSE since 1989, focusing on human risk, diagnostic tests and the treatment of any disease condition that may appear in humans as a result of BSE.**

Table 1. vCJD in UK

Year	1995	1996	1997	1998	1999	2000	2001
Cases	3	10	10	18	15	28	38*

*Predicted for 2001 at current rates.

Table 2. European BSE

Country	1999	2000	2001*	Country	1999	2000	2001*
Belgium	3	9	22	Greece	0	0	1
Czech Rep.	0	0	2	Italy	0	0	23
Denmark	0	1	3	Netherlands	2	2	12
France	31	162	140	Portugal	170	163	44
Germany	0	7	94	Spain	0	2	57

*Until August, if known.