

Britain: New wave of human BSE/vCJD feared

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News reports have raised the spectre of a second wave of deaths in Britain from variant Creutzfeldt-Jakob Disease vCJD as a result of people, overwhelmingly young adults, having consumed meat or meat products from cattle infected with Bovine Spongiform Encephalopathy (BSE), or Mad Cow Disease.

The first case of BSE was in Britain in 1984. It was associated with the growing practice of recycling the corpses of dead farm animals in the production of protein cattle feed. A similar disease in sheep had been recognised for many years. It is thought that the prion agent which causes the disease in sheep, Scrapie, had been able to jump the species barrier and infect cattle as well—the result of recycling animal remains in the production of cattle protein pellets.

The British Conservative government of the time strenuously denied the possibility of the disease passing to human beings. However, in 1992, 18-year-old Stephen Churchill became the first to be diagnosed with the human form of the disease. He died in 1995, and in 1996 the Tory government was forced to admit humans could become infected as a result of BSE in cattle.

BSE and its human form are a result of the action of prion protein. Prions occur naturally in the body, but there exists an abnormal form which has a different shape. If this abnormal form is introduced into the body of an animal or human, such as by ingestion, it can cause other normal prions to change shape in a kind of chain reaction process. This causes the prion protein molecules to clump in the brain and leads to holes developing in the brain tissue. The result is ever increasing loss of brain function leading to death.

The majority of cases of the disease have occurred in Britain. Over 160 have now died from vCJD. Outside of Britain, there have been around 20 deaths in France and a handful of cases in other countries. The number in Britain dying from the disease peaked in the year 2000 when 28 cases were recorded. The last few years have seen the number of deaths decline to single digits.

Prion protein production in the bodies of humans and animals is controlled by a particular gene. To date those humans that have died from vCJD have had a particular genetic makeup. Such gene types are classed as MM, comprising up to 42 percent of the population, with 47 percent of the population having so-called MV genes and the remaining 11 percent having what is called a VV genetic makeup.

The current news reports concern a young man diagnosed as having vCJD but with genes of the MV type. If the diagnosis of vCJD is confirmed (this can only be done by invasive procedures such as brain biopsy) his will be the first case to occur outside the MM genetic group.

Interviewed on the BBC *Newsnight* programme, the chairman of the government's Spongiform Encephalopathy Advisory Committee (SEAC), Professor Chris Higgins, said if this new case was confirmed to be from the MV genetic cohort, it could signal a new wave of cases. He estimated the numbers dying could be between 50 and 350.

The programme made the point that in Britain most of the adult population was exposed to the disease, through the food chain. Professor John Collinge of the Institute of Neurology has stated that around one in a thousand may be silent carriers of the disease. This poses a possible

danger of transferring the disease by blood transfusion.

As yet there is no test to screen blood for the disease, although scientists are working on one and it is hoped that it will be available by the end of next year.

Newsnight also interviewed Professor Hugh Pennington, Emeritus Professor of Bacteriology at Aberdeen University. He said he was not surprised that a possible diagnosis of the disease had been found in someone outside the MM cohort. A similar situation happened in the case of Kuru, a brain-wasting disease found in Papua New Guinea, which resulted from the custom of native people eating body parts of tribal members who had died.

With Kuru, people with the MV genetic makeup had developed the disease many years after the initial exposure. Once the connection was made between the disease and the practice of eating human remains, it was stopped. However, cases of Kuru still occur. The MV cohort has a longer incubation period for the disease. Professor Pennington thought that we could be seeing the same thing happening with vCJD.

The January edition of *The Lancet* medical magazine poses the possibility that "a second wave of CJD with a longer incubation time might hit these shores, but we do not know whether this will be a tidal wave or just an imperceptible ripple."

The fact that a group of the population may have a longer incubation period for the disease does not necessarily mean that they might be less susceptible to the disease. The main *Lancet* article by Simon Mead et al on the genetic factor and the occurrence of vCJD makes the point: "Whether these effects are on the incubation period rather than susceptibility, such that further waves of BSE-associated prion disease with longer incubation periods might occur in the years ahead is unknown."

Also appearing on the *Newsnight* programme was freelance journalist Christine Lord. Her son, Andrew Black, died one year ago at the age of 24 from vCJD. She is making an appeal for people to be made aware of a possible second wave of the disease and is campaigning to expose politicians and advisors in previous government administrations who were aware of the dangers and covered them up. She holds them responsible for the death of her son.

A previous BBC programme, *Inside Out*, included footage of the last months of her son's life as the disease took hold. It also included footage of her interviewing Professor John Collinge, who explained that he had been subjected to intimidation by government departments at the time of the BSE crisis and ordered not to speak out on what was taking place.

Professor Richard Lacey of Leeds University did speak out and was ridiculed and vilified by the government of the time.

Scientists have expressed concern that research into vCJD could be cut back. Professor Higgins, in the *Observer* newspaper in August, before the current development, stated, "We face at least one more wave of variant CJD in Britain. That suggests we need to maintain our research efforts into finding treatments for the condition."

In the same article, Professor Collinge explained, "So far, UK funding has remained strong in its support for CJD work, though researchers in France and Germany have already noted grants for CJD work are drying up. That would be a mistake if it were repeated here."

There is as yet no cure for this devastating disease and the current funding of research can easily come under threat as the economic crisis deepens.



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