Britain: Thousands more people could be infected with vCJD

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The *Journal of Pathology* published a report on the Internet on May 20, which reveals that thousands of people in Britain could be carrying variant Creutzfeldt-Jakob Disease (CJD)—the human form of Bovine Spongiform Encephalopathy (BSE). BSE, or "Mad-Cow disease", is a brain-wasting disease that spread through British cattle in the late 1980s and 1990s.

The cause of the disease is thought to be a misshapen form of a protein known as a prion that occurs naturally in the body. The misshapen form of the prion is passed to humans by the ingestion of meat infected with BSE. It causes normally shaped prion molecules to deform in a chain reaction. These deformed prions clump together and produce holes in the brain, leading to increasing loss of neural function and eventually to death.

The disease was spread to humans by the consumption of infected beef, despite claims from the British government at the time that this was not the case. Since 1995, 141 mainly young people have died from the condition and a further six cases have been diagnosed.

The recent *Journal of Pathology* report was based on research by David Hilton from the Department of Histopathology at Derriford Hospital Plymouth; Azra Ghani, Department of Infectious Diseases Epidemiology in the Faculty of Medicine at Imperial College London; James Ironside, National CJD Surveillance Unite at the University of Edinburgh; and other medical scientists.

They studied tissue samples anonymously taken from over 12,500 appendices and tonsils that were removed in routine operations. These organs have been shown to be sites of accumulation of prion proteins prior to clinical onset of the disease. Of these samples, three contained traces of the deformed prions that cause vCJD. If projected onto the population as a whole this figure would give an estimate of around 4,000 people currently incubating the disease.

Professor John Collinge, head of the Medical Research Council Prion Unit at St Mary's Hospital in London, said of the report, "I find these results very concerning. Our experience is that looking at appendix samples will underestimate the true picture. In addition, no test is going to be 100 percent effective, and you don't know at what stage in the incubation period the test will be positive."

In two of the infected samples the pattern of accumulation of the prions was unlike those seen in tissue from people actually diagnosed as suffering from vCJD. Yet the accumulation of such prions is considered a unique indicator of vCJD. It is thought the new pattern may represent some form of carrier version of the disease, which is present in the body but stays at a sub-clinical level.

This could represent a real danger if such carriers gave blood or underwent surgery. The prion particles are known to be very resistant to destruction and can survive the normal sterilisation processes carried out on surgical equipment. Professor Collinge called on the government to reconsider the use of disposable surgical instruments in cases where there may be the risk of transmission, such as the removal of tonsils.

Professor James Ironside, who heads the CJD surveillance unit at Edinburgh and who participated in the study, said:

"There would seem to be more positives than you would expect. That may be because of genetic differences and susceptibility, but it may also be that you can have a sub-clinical infection that never progresses to produce symptoms—a 'carrier state'. These findings do have to be taken seriously. Generally one has to be cautious about interpreting these data, but

they may indicate that there are people who are not infected in the normal way but could represent a source of infection."

The Health Protection Agency, successor organisation to the Public Health Service, had recently reported that the number of deaths from vCJD had peaked. But many scientists still fear a bigger epidemic could take place. All of the victims so far have had a particular genetic pattern. There is concern that these may be the ones to show rapid susceptibility to the disease but that others may fall victim to the disease over a longer period of time.

Kuru, a type of spongiform disease similar to vCJD, was prevalent in some tribal peoples in Papua New Guinea. It was discovered to result from the practice of eating body parts, particularly brains, from deceased relatives as part of a religious ceremony. In this disease the incubation period could be measured in decades. In the case of human BSE the normal incubation period may be longer, given that the disease has jumped the species barrier.

The government has announced a more extensive investigation, involving the study of around 100,000 tonsils and appendices over a three-year period.

In a comment to the BBC, Don Simms, whose son Jonathan is suffering from vCJD, said, "The fact is we just don't know how many people are affected. We need to find out."

Simms has been to court to win the right for Jonathon to be treated with a drug that, although had been shown to be effective in halting the progress of the disease, had not undergone clinical trials. To date the drug does seem to be effective in halting the progression of the disease.

Frances Hall, whose son Peter died of vCJD in 1996, speaks on behalf of the families of victims of the disease. Commenting on the recently published report she told the *World Socialist Web Site*, "It is not good news. It's 10 years on and there are still more questions than answers. We urgently need a reliable blood test so that human-to-human infection can be eliminated. The government insists that blood transfusions are safe now, but at the same time anyone who has had a transfusion is banned from donating blood. Also babies born after 1996 can only receive blood, which is sourced in countries where there is no BSE.

"The truth is that we do not know how many individuals are incubating the disease. The estimate was 500 and now is 4,000, but it could go even higher."

Hall said that the families of vCJD victims try to support one another. Every year on March 20 they gather in London by the river Thames opposite parliament and read out the names of the dead and throw a white rose for each one into the water as a commemoration. She added, "My feeling on that day is to shake a fist across at the House of Commons. For us March 20, 1996, was a 'red letter day' because it was on that day that the government finally acknowledged what we all knew, that there was a link between BSE and vCJD."

Human BSE represents a potential epidemic time bomb, for which the necessary resources and expertise should be mobilised to develop accurate figures on those likely to be affected and for urgent progress to find a cure.