Significant breakthrough in diagnosis of human BSE

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The British Labour government may be forced to carry out mass screening for nvCJD (new variant Creutzfeldt-Jakob Disease), the human form of BSE (Bovine Spongiform Encephalopathy) or mad cow disease, which became endemic in the country's cattle herd during the 1980s. Twenty-seven people in Britain have now died from nvCJD, which progressively destroys brain function leaving the patient blind, mute and incapacitated.

The prospect of mass screening follows publication of a research letter in the August 29 issue of the medical journal, *The Lancet.* The article reported the work of Dr John Zajicek and his team at Derriford Hospital, Plymouth, where a patient, Tony Barrett, had died from nvCJD.

Barrett, a coast guard, first developed symptoms of human BSE in May 1996 and died in June this year. In September 1995, eight months before the onset of the disease, Barrett underwent an operation to remove his appendix at Torbay Hospital in Devon. After his death, doctors examined his appendix which had been routinely stored by the hospital, and found evidence of PrP (prion proteins, the infectious agent for the disease) in the lymphoid tissue. They used a procedure developed by scientists in Holland 20 years ago, which detects scrapie (another Transmissible Spongiform Encephalopathy) in the tonsils of sheep aged over 10 months.

The Lancet article points out that these findings are the first demonstration of PrP in tissue in humans during the incubation period of nvCJD. It also points out that the involvement of tissue in the gut, before clinical onset of the disease, is in keeping with an enteric route of entry for the disease agent--i.e., through the consumption of infected food.

This discovery is extremely significant, as it means that tests can be carried out before clinical symptoms of nvCJD are displayed. Until now confirmation of the disease could only be made after a post-mortem examination of the victim's brain.

Hospitals routinely collect sample tissues from the 44,000 appendectomies and 800,000 tonsillectomies carried out each year. *The Lancet* has urged the government to carry out a large scale-screening programme of all hospital specimens dating from the onset of the BSE epidemic in cattle. This would provide data on the number of people incubating the infectious agent and at risk of developing the disease. The figures would still not be conclusive, however, as it is not known at what stage of the incubation period the lymphoid tissue becomes involved, and whether this inevitably results in the development of nvCJD.

Following publication of these findings Sir Kenneth Calman, the government's chief medical officer held a meeting with Dr James Ironside of the Edinburgh CJD Surveillance Unit and representatives from Derriford hospital. Calman stated that there would be no immediate change in health controls, but that tests would be performed on other appendix samples, 'to see what it means'.

Ministers have subsequently approved a review of thousands of relevant laboratory specimens and have stated that should one more case be identified, hospitals will begin to screen all patients awaiting tonsil or appendix operations.

Dr Stephen Dealler, a scientist working in the field, told the *World Socialist Web Site* that it has been known since 1997 that the test used by Dr Zajicek was effective in detecting nvCJD. Yet there has been no explanation of why a similar investigation had not been proposed earlier by the government Spongiform Encephalopathy Advisory Committee (SEAC). It raises

the danger that similar work has been undertaken by government scientists and not reported.

It is known that the disease can be passed on through blood, and at least one human BSE victim is known to have been a blood donor. Dr Dealler raised the possible contamination of blood supplies four years ago, but regulations were not changed until earlier this year. The use of white blood cells from British blood donors is now banned. Instead, supplies are imported from abroad.

The publication of the research letter in *The Lancet* raises several immediate issues.

Surgical instruments used on known, or even suspected, human BSE patients must be destroyed, as the infectious agent is resistant to sterilisation. As Tony Barrett was not displaying symptoms of nvCJD at the time of his appendix removal, it is likely that the surgical apparatus used during his operation will have been used in other procedures, posing the risk of cross-contamination. Similar dangers are present with other operations like corneal transplants.

More broadly, Labour's approach to BSE and its impact on human health has been a continuation of that taken by the previous Tory government. They have claimed that the danger is minimal and has largely passed. Now the possibility exists of determining the incidence of nvCJD in the human population. It is known that approximately half the British population has a genetic make-up that makes them susceptible to infection.

This has posed the dilemma of how to proceed in the case of positive testing. Human BSE is a disease for which there is as yet no known cure. Diagnosis is the equivalent of a death sentence. In addition, fears have been voiced that those incubating the disease could find themselves in a 'legal limbo'--prevented from taking out insurance policies and bank loans--and turned into social pariahs. Yet the withholding of a positive result would further endanger public health. Researchers have already begun preparing procedures and ethical rules for the initial investigations, to be funded by the Medical Research Council.

These problems notwithstanding, the ability to screen for nvCJD is an important breakthrough. It must not be allowed to be sacrificed on the altar of economic and political expediency as has happened all too often in the history of this terrible disease.

See Also:
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An exchange of letters on the Mad Cow Disease (BSE) crisis
[23 July 1998]