

SUMMARY OF THE FINAL REPORT OF THE INVESTIGATION INTO THE NORTH LEICESTERSHIRE CLUSTER OF VARIANT CREUTZFELDT-JAKOB DISEASE

The investigation was carried out by Dr Gerry Bryant and Dr Philip Monk who have prepared and present this report to Leicestershire Health Authority.

1. INTRODUCTION

The cluster of variant Creutzfeld-Jakob disease in the North Leicestershire area was first recognised in July 2000. Between August 1996 and January 1999 five people developed symptoms that were later recognised as being those of variant Creutzfeld-Jakob disease and they have all died. They all lived in the Wreake and Soar Valley area in North Leicestershire from 1980 until 1991.

The investigation concentrated on the period from 1980 to 1991, as this was the only time period when a common exposure could have occurred.

2. INITIAL FIELDWORK

At least one relative of the people with variant Creutzfeld-Jakob disease was interviewed to examine possible exposures to the BSE (Bovine Spongiform Encephalopathy) Agent.

A questionnaire study was carried out with parents of children at Queniborough Primary School to examine their purchase and consumption of food when they were children themselves during the 1980s.

This was followed by a questionnaire study of people living in the village of Queniborough. The questionnaire asked those people who were parents of children of similar age to the cases about their purchase and consumption of meat during the 1980s.

Butchers, farmers, auctioneers and others involved in the meat trade were interviewed to build a picture of the dairy and beef industries during the 1980s.

3. RESULTS FROM THIS INITIAL FIELDWORK

1. Possible Exposures

A number of possible risk factors were unlikely to provide an explanation for a link between the people with variant Creutzfeld-Jakob disease. They were: -

- Surgery and blood transfusions
- Dental surgery
- Occupational exposure
- Immunisations
- Injections, body piercing, cuts and animal bites
- Baby foods, school meals and drinking water
- Manganese (data from the British Geological Survey showed that the area of Leicestershire does not have a high level of Manganese in the soil)

1. INVESTIGATION INTO FARMING PRACTICE

1. *Cattle raising practice*

Local beef cattle were raised alongside dairy cattle. This meant that beef cattle were fed meat and bone meal supplements from the age of 6 days rather than 6 months, which is the case for pure beef herds. They therefore had a greater lifetime exposure to the BSE agent in meat and bone meal than cattle who did not receive meat and bone meal until the age of 6 months.

The area of Leicestershire that supplied beef cattle to the local food trade had a moderately high incidence for BSE meaning that a number of cattle across a number of farms had the disease.

At the beginning of the 1980s, local beef cattle were a by-product of the dairy industry and were usually Friesian Hereford crossbred cattle. Such cattle were usually slaughtered between 30 and 36 months of age because they are slower to fatten.

The average age for onset of BSE in cattle was between 4 and 5 years. However, the BSE Inquiry notes that although the numbers were small, there were a few cases in which clinical onset occurred between 20 and 30 months. The youngest animal in England being just 20 months old. This means that the older Friesian cross bred cattle used in the meat trade in Leicestershire were more likely to have sub-clinical BSE infection and to be infectious. Back calculations from the BSE epidemic suggest that British cattle must have had BSE from the mid 1970s onwards. This area of Leicestershire reported BSE as soon as it became notifiable in 1988 which means that some cattle were likely to have had BSE during the period that we were investigating.

2. *Cattle slaughtering practice*

In both large and small abattoirs, cattle were slaughtered using a captive bolt. In the local abattoirs and butchers who slaughtered, a pithing rod was also used. (see appendix – terminology) In small abattoirs the carcass was wiped down with a cloth to remove unwanted tissue. In large abattoirs the carcass would be hosed down. In the early 1980s there was no legal requirement to hose down a carcass. Skilled butchers reported that hosing a carcass down would make the meat go 'sour'. The practice of wiping a carcass with a cloth meant that there was a possibility of cross contamination of meat with brain and nervous tissue in those butchers who removed the brain from a beast's head.

3. *Carcass purchase*

Most butchers in the area bought meat from wholesale suppliers. A small number would select cattle at Melton Market or directly from a local farm for slaughter either by themselves or in a small nearby abattoir.

Wholesale meat suppliers purchased carcasses from a number of abattoirs that in turn selected cattle from a number of cattle auctions.

2. RESULTS OF INVESTIGATION INTO BUTCHERING AND MEAT PROCESSING

1. *Butchering practice*

1. Whole carcass processing

A small number of butchers either slaughtered beasts in their own small abattoirs or had beasts slaughtered by a nearby small abattoir. The butcher then processed the whole carcass. For those butchers who had a market for brain, they removed it from the beast during the process of recovery of head meat. The rest of the carcass was then boned and jointed. The removal of brain meant that there was the possibility that other meat could be contaminated with brain material. Brain contains the BSE agent and is therefore potentially infectious. During the 1980s, this process was legal and represented traditional butchering practice. It was decreasing because of a declining consumer market for brain. In the past and in particular during the war years, brain had been seen as an excellent source of protein.

2. Wholesale purchase

By the beginning of the 1980s many butchers had moved to purchasing either sides of beef, quarters of beef or vacuum packed pre-prepared cuts of beef rather than whole carcasses. A small number of these butchers would also purchase heads in order to remove the tongues to prepare for pressed tongue and sometimes the cheek meat usually for pet food. It was very rare for such butchers to remove the brain, as by this time there was often no market for brain.

3. THE HYPOTHESIS

Our initial work suggested that there was an association between the cases of variant Creutzfeldt-Jakob Disease and the consumption of beef purchased from butchers where there was a risk of 'cross-contamination' of beef carcass meat with bovine brain.

The essential elements of our hypothesis are that:-

- the beasts used were locally reared cattle
- the beef cattle were a by-product of the beef industry and therefore fed meat and bone meal from day 6 onwards giving them a greater lifetime exposure to feedstuff that was potentially contaminated with the BSE agent
- they were predominantly Friesian crossbred cattle which were slow to fatten and therefore slaughtered at close to three years
- they were slaughtered in small abattoirs which employed pithing and without the washing down of the carcass
- the heads were split to remove the brain
- during brain removal, if the meninges (the membrane that covers the brain) are broken,

because brain is of a gelatinous consistency, when handled, it then has a tendency to be adherent

- carcasses were wiped with cloths increasing the risk of cross-contamination

1. TESTING THE HYPOTHESIS

Our study to test the hypothesis was carried out with the approval of the Leicestershire Research Ethics Committee.

A relative of each case variant Creutzfeld-Jakob Disease in the cluster was re-interviewed using a structured questionnaire. The questionnaire asked about dietary history including meat consumption and from where meat was purchased during the period 1980 to 1991. A relative of each of thirty age-matched controls, six for each case, was interviewed using the same questionnaire. An attempt was made to interview all butchers, supermarkets and freezer food centres identified by the controls to ascertain whether they or their suppliers used cattle heads and removed bovine brain, thus creating the opportunity for cross-contamination.

2. RESULTS OF THE CASE CONTROL STUDY

1. *Butchers, supermarkets and freezer food centres used by cases and controls*

Four of the people who developed variant Creutzfeld-Jakob Disease bought and consumed beef from one of two butchers during the early 1980s. One of these butchers slaughtered beasts in his own abattoir. This butcher normally processed three beasts a week. He ceased trading in 1989. The other butcher had beasts slaughtered in a small nearby abattoir. He processed four to five beasts a week. This butcher's business ceased trading in December 1982. It has not been possible to trace the butcher who was used regularly by one family during the first half of the 1980s. It is unlikely that he removed brains or even purchased the heads of beasts. He did not slaughter beasts himself or use a small local abattoir.

People acting as controls used a total of twenty butchers, four freezer food centres and seven supermarkets. With the exception of one butcher who supplied one of the controls, all other outlets were traced and staff interviewed to ask whether they removed the brain. Three butchers used by controls removed brains. One of these butchers was also used by one of the people who developed variant Creutzfeld-Jakob Disease. With the exception of this one butcher and one other who also split the heads, the butchers used by the controls processed between one and two sides of beef or less a week. A side of beef is half of the carcass. We were able to trace both butchers and buyers who worked for the supermarkets used by people in this study. None of them reported the use of head meat during the 1980s. Sides of meat or vacuum packs were purchased from wholesalers. The wholesalers who supplied the supermarkets and freezer centres did not split heads to remove the brains. The skulls were sent either to specialist head boning plants or to renderers after removal of the head meat. The skulls were never split.

2. Interview results

The study showed that the relatives reported that the people with variant Creutzfeld-Jakob Disease were 15 times more likely to have purchased and consumed beef from a butcher who removed the brain from a beast compared with controls who purchased meat from outlets where cross contamination with brain material was not a risk. This result is statistically significant and is therefore very unlikely to be a chance finding. ($p = 0.0058$ and the 95% confidence interval is 1.6 to 138.9)

3. LIMITATIONS OF THE STUDY

A number of factors may have influenced the result.

Our work suggests that family size, age and education of children of controls were representative of people living in the area meaning that our controls who were randomly selected are likely to be representative of people living in the area.

It is possible that we were in some way able to influence the outcome of the study by the way in which we wrote down the answers to the questions that we asked both of the cases and controls. Wherever possible we interviewed people together to ensure that the interview technique was the same. All interviews were recorded on tape. Whilst we knew the hypothesis that was being tested, at the time of the interviews we had no knowledge of the butchering practices of most of the meat suppliers identified by the controls. These butchers were interviewed after the interviews with controls had been completed.

People may not be able to remember what they gave their families to eat twenty years ago. However, our hypothesis was dependent on where people shopped and we were not attempting to identify every single item of food that was eaten. Rather we explored the usual patterns of meat consumption and the usual sources where beef was purchased.

4. CONCLUSIONS FROM THE STUDY

We have found an association which provides a biologically plausible explanation suggesting that four out of the five people with variant Creutzfeld-Jakob Disease may have been exposed to the BSE agent through the purchase and consumption of beef from a butcher's shop where the meat could be contaminated with brain tissue. On a national basis, it is unlikely to explain how all of the people who have developed this disease were exposed to the BSE agent.

Assuming that we are correct in our explanation, we have shown that for one of the butchers, the exposure took place before December 1982. For the other the risk of the exposure continued until that butcher ceased trading in 1989. Analysis of the exposure of our cases to this butchering practice points to an incubation period for the development of variant Creutzfeld-Jakob Disease of between ten and sixteen years. This is the first time that it has been possible to provide an estimate of the incubation period.

We have shown that it is possible to examine by traditional epidemiological methods

exposures that took place twenty years ago.

5. RECOMMENDATIONS

The explanation that we have found needs to be tested for other people who have died of variant Creutzfeld-Jakob Disease.

Surveillance for variant Creutzfeld-Jakob Disease needs to be maintained in Leicestershire.

6. THANKS

We would like to pay tribute to the courage of the families of those people who died of variant Creutzfeld-Jakob Disease who through their willingness to relive their pain have helped us to learn more about the way in which this disease spread from animals to people.

We thank all of those who willingly gave of their time to be interviewed as controls together with the butchers, farmers, auctioneers and others in the food trade who helped us with our study.

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Gerry Bryant and Philip Monk
Leicestershire Health Authority 17 March 2001

BACKGROUND INFORMATION

INVESTIGATION OF VARIANT CREUTZFELD-JAKOB DISEASE

All people with variant Creutzfeld-Jakob disease are reported to the National Creutzfeld-Jakob Disease Surveillance Unit, which is based in Edinburgh. Doctors from the unit visit all people with the disease. A detailed history is taken and the diagnosis reviewed.

MEAT AND BONE MEAL

Meat and bone meal is a protein supplement fed to cattle. It was made from rendering the parts of sheep, pigs, chickens and cattle that were not consumed by people.

As little as 1 gram of infected material when fed to cattle is known to cause BSE in 70% of those animals fed the infected material.

In December 1988 it was recognised that the likely way in which BSE was spread was through feeding cattle protein supplements containing meat and bone meal. In November 1989, through a voluntary ban, Animal food manufacturers stopped the inclusion of any Specified Bovine Offal in Meat and Bone Meal fed to ruminants. This voluntary ban was made law in September 1990 in the Bovine Spongiform Encephalopathy (No 2) Amendment Order 1990.

TERMINOLOGY

BSE Bovine Spongiform Encephalopathy

Captive bolt stunner

An instrument for stunning animals before slaughter, powered by a cartridge or compressed air which drives a bolt out of a barrel for some four inches and then retracts it into the barrel.

Pithing

Insertion of a rod through the stun hole in the head of cattle to prevent the animal kicking (a reflex action which sometimes occurs after stunning)