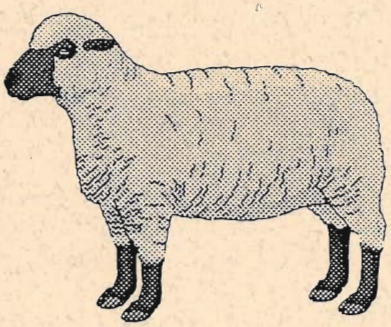
MAD COWS

AND

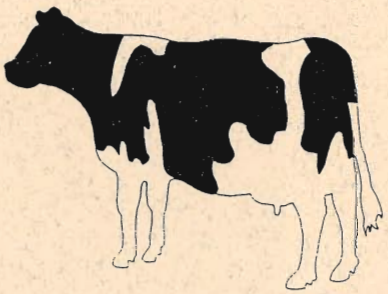
ENGLISHMEN

TRANSMISSIBLE SPONGIFORM ENCEPHALOPATHIES

SCRAPIE



BSE



CJD

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RESPONSIBILITIES AND RIGHTS

REPORT PREPARED BY:

ROSIE DWYER

KELLOGG RURAL LEADERSHIP COURSE 1997

ABSTRACT

Transmissible Spongiform Encephalopathies (TSE's) are a group of diseases that affect certain types of animals and can also be found in humans. Transmissible refers to the fact that the diseases can be transferred or spread. Spongiform refers to the groups of holes which form in the brain and Encephalopathy means degenerative brain disease.

Each TSE disease has a separate name but all have the same underlying infective agent - the Prion. The diseases are characterized by a long incubation period; degenerative changes in the brain associated with a `prion' agent; and all are invariably fatal.

Dr Stanley Prusiner labeled the unusual infective agent as a Prion. The term links it with the proteinacious infective properties that it has. Prions are not bacteria or a virus but protein molecules that are capable of replication through changing the shape of normal proteins. The normal prion, found in mammals, is coded for by a gene found on chromosome 20 also known as the PrP gene. The abnormal form of the PrP gene is coded as PrPsc.

There are four prion diseases that occur in humans: Creutzfeldt Jakob disease (CJD), Gerstmann-Straussler-Scheinker syndrome; Kuru; and Fatal Familial Insomnia. There are also four TSE diseases which occur in animals: Scrapie of sheep and goats; mink TSE; chronic wasting disease of American mule deer and elk; and Bovine Spongiform Encephalopathy (BSE) - also known as `Mad Cow Disease'.

Scrapie in sheep and goats is the most well known TSE and has been around for almost three centuries. It is found in Britain, Europe and North America. The characteristics of the disease are loss of condition, nervous signs and a peculiar skin irritation that causes the sheep to rub or scrape themselves to relieve itching - hence the term Scrapie.

Bovine Spongiform Encephalopathy or Mad Cow Disease appeared in the United Kingdom in Kent in 1986. It was common practice for farmers to feed their cattle Meat and Bone meal - supplementary feed that comprised ground up waste material from sheep and beef. The way in which the meat and bone meal was processed changed in the early 1980's, and this has supposedly led to infective scrapie tissue being fed to cattle. The previous method of rendering the meal inactivated the infective scrapie tissue - which is why it had not appeared before this process changed.

Subsequently a ban was imposed on feeding ruminant waste to ruminants and a ban was placed on Specified Bovine Offals being allowed in to human food production in July 1988.

The number of cases of BSE have not fallen off as sharply as the British Ministry of Agriculture, Fisheries and Food (MAFF)

had hoped for. However, due to the afore mentioned bans and a cull of cattle over 30 months of age, the numbers are expected to tail off by the year 2000.

Mark Purdey , a UK organic farmer has a theory that the Government enforced use of organophosphates to kill the warble fly, has led to the out break of BSE. His controversial theory has not made him many friends within MAFF and chemical companies who supplied the treatment. He and his co-workers have had some unfortunate accidents in their quest for the truth of this matter.

Compensation totaling millions of pounds has been paid out to farmers that have had cattle affected by BSE.

The UK Government has been condemned in the way in which they handled the BSE epidemic. Some would say with an irresponsible attitude. Up until March 1996, the Minister of Agriculture had consistently denied that there was any risk to humans in eating beef affected by BSE (contrary to what scientist's had been trying to tell them). Then in March 1996, the UK Ministry of Health announced that 10 cases of a new variant strain of CJD had been isolated and there were tentative links to the cause of the disease being the BSE agent.

Currently, the European Union has banned all British beef and beef product imports. This ban also extends to tallow which affects New Zealand tallow exports to China. The European Union does not accept that New Zealand is BSE or scrapie free - they insist that we can not prove that we don't have either at present.

Creutzfeldt Jakob Disease occurs in most countries at a rate of between 0 and 5 cases per million people.

The new variant strain of Creutzfeldt Jakob Disease is causing concern in that it is targeting predominantly young people (18-40 age group) it has a long incubation period, up to 24 month illness duration and more importantly is linked to BSE as the causative agent.

There are many on going research projects and studies being done in relation to transmissible spongiform encephalopathies. Questions that arise in the search for more answers are:

what are the possible methods of transmission between species ?
why is new variant CJD only targeting young people ?
is there proof that BSE is the cause of new variant CJD ?
is it safe to be eating British beef ?
what are the chances of the new variant CJD becoming an epidemic in the UK ?

This report provides an overview in to the current information available on scrapie, BSE, CJD and new variant CJD.

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1. INTRODUCTION

This report reviews the data and information currently available on Transmissible Spongiform Encephalopathies - specifically Scrapie, Bovine Spongiform Encephalopathy (BSE) and Creutzfeldt-Jakob Disease (CJD).

Knowledge about these group of diseases is not complete. Scrapie has been around for nearly three hundred years but it is not known what causes the disease or whether it has ever been transmitted to man. There is a belief held that scrapie was the cause of the BSE epidemic in the United Kingdom within the last 10 or so years.

BSE or 'Mad Cow Disease' as it is also called, has cost the United Kingdom cattle industry millions of pounds and with the appearance of a new form of Creutzfeldt-Jakob disease, there are links being made between it and BSE. This introduces fresh alarm regarding the safety of British beef and beef products.

The British Government's response to the BSE epidemic was to provide compensation to farmers that had lost stock to the disease. The Government was, however, slow to heed the warnings from scientists and researchers about the risk to man from BSE. Not until March 1996 when the first ten cases of the new variant from of CJD were reported, did the Government issue a statement that BSE and the new variant CJD may be linked. While the evidence is not conclusive, current information available points to there being a link.

While steps were taken to curb the BSE epidemic, policing these measures has been a difficult task. The ban on ruminant feed and Specified Bovine Offals was not as effective as UK MAFF had hoped probably due to non-compliance of farmers and the fact that cattle born after the bans had contracted BSE.

There appears to be a relationship between scrapie, BSE and CJD. Scrapie infected meat and bone meal is thought to have caused BSE, and BSE infected beef products is thought to be the cause of the new variant CJD.

The completion of research and experiments into the area of transmission between species may give more definitive answers.

The issues of animal and food safety rests with all concerned in the meat industry chain. These issues are not limited to the UK alone. In New Zealand we must be diligent about animal health, production safety and food safety to the consumer. The consumer also has a right to know what they are purchasing and eating.

Producer - Processor - Manufacturer - Consumer and Government must all take part in the issue of safety.

The information in this report is current up to November 1997.

TRANSMISSIBLE SPONGIFORM ENCEPHALOPATHIES

2.1 SCRAPIE

Scrapie has been categorized as a slow virus disease. It is a fatal transmissible disease found in most breeds of sheep and can affect either sex, although usually seen in breeding ewes. The disease is characterized by a long incubation period of up to 8 years with an average incubation of 3.5 years.

While there are no gross pathological lesions to be noted, there are changes within the minute structure of tissues. Specifically, within the central nervous system vacuoles or spaces appear within the cells creating a spongy-like lesion.

Scrapie manifests itself within the sheep and is exhibited in two ways.

Firstly, Pruritis or irritation of the skin. This causes the sheep to rub or 'scrape' against fixed objects which in turn causes extensive loss of wool.

Secondly are neurological signs such as exaggerated and uncoordinated gait. A high stepping gait in the front legs is common with failure of muscle co-ordination in the hind legs. At rest a sheep may sway, stand awkwardly or even lose its balance. The progression of the disease leads to loss of condition and emaciation. (Bruere & West 1993)

Scrapie produces a peculiar skin irritation followed by rubbing, nervous signs and severe loss of condition.

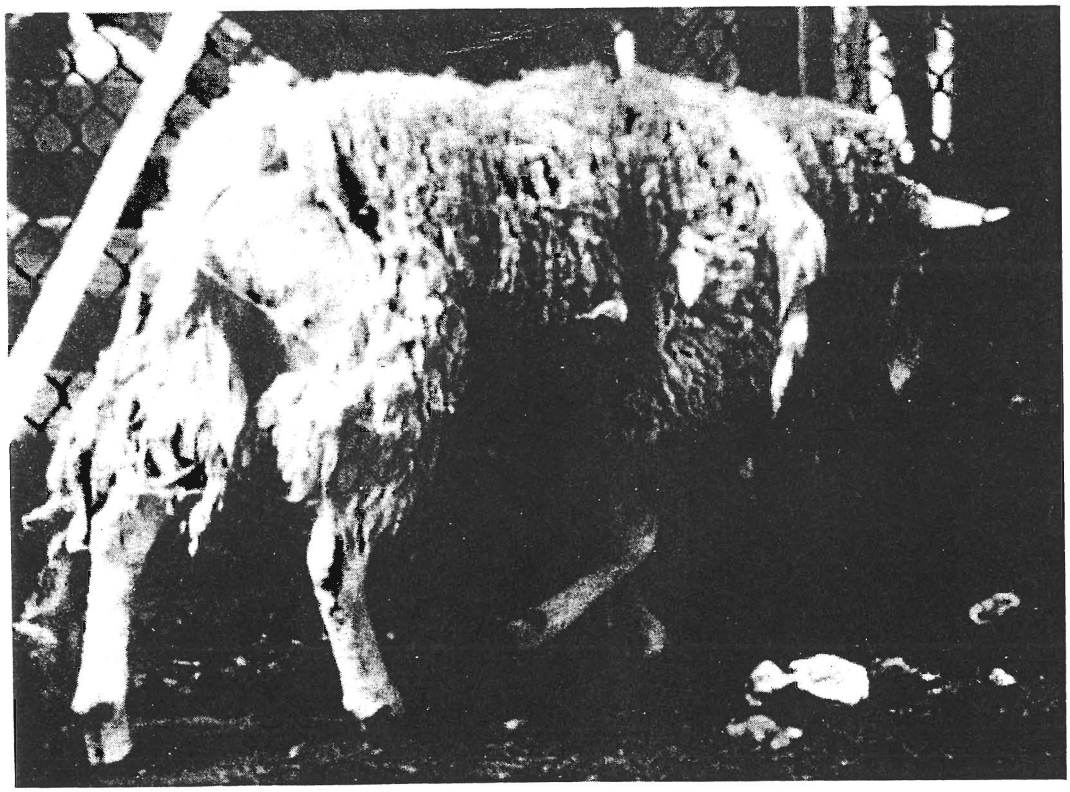


Photo 1a. Sheep affected with advance scrapie
(Sheep Production vol 2: p269)

2.2 BOVINE SPONGIFORM ENCEPHALOPATHY

"Mad Cow Disease"

Much has been written about Mad Cow Disease since its discovery in the 1980's. Because it is relatively new , it has had the full benefit of the media to tell its story - a luxury not afforded its cousin scrapie, which has been recorded for 250 years. Television, radio, newspapers, books and magazines have all played a part in dispersing information, statistics and stories surrounding the out break of BSE.

In 1986, BSE was confirmed in dairy cows on a farm in Kent, England. Their irritable behaviour, fear of the farmer, changes in temperament and subsequently loss of appetite and milk production preceded the cows deaths. The brains from the cows were analysed by the Central Veterinary Laboratory in Weybridge, U.K. and the spongiform encephalopathy was diagnosed. The disease has probably existed since 1982. It is a disease almost certainly linked to scrapie, possibly through the feeding of scrapie infected meat and bone meal to cattle.

In Britain meat and bone meal had been fed to cattle for many years prior to the out break of BSE. Bruere & West(1993) and Science behind the News (June 1996) both report on the meat and bone meal theory. In 1981 manufacturing plants changed the way in which they processed animal carcasses into meat and bone meal. Prior to this time the plants had used a hydrocarbon solvent and heat treatment to remove fat from the meal, which appears to have prevented the scrapie agent from entering the meal. However, removal of these processes no longer protected cattle from the scrapie agent.

Another theory is that it was BSE that was in meat and bone meal and not scrapie. The change in manufacturing of the meal allowed BSE to spread amongst cattle. Why BSE hadn't appeared before this time seems to be a case of good luck rather than good management. BSE may have been around as a rare cattle disease with similar symptoms to that of grass staggers. By chance around 1980's an infected cow's waste tissues were used in meat and bone meal and fed to other cows and so on.

The processing changes occurred from the late 1970's until 1982. BSE was not diagnosed until 1986 though it possibly existed since 1982.

> "the 4-year lag between when the solvent was removed from the meat and bone meal manufacturing process and the first reported cases of BSE in 1986 suggested that the disease had a long incubation period. This was later proved correct."
>
> Science behind the News 1997

In the period between 1981 and 1986 infected cattle showed no signs or symptoms of the disease. Some of these unsuspected cattle would have been used in meat and bone meal - the epidemic was possibly driven by the 'recycling' of infected cattle.

2.3 HUMAN SPONGIFORM ENCEPHALOPATHIES

CREUTZFELDT-JAKOB DISEASE

Creutzfeldt-Jakob Disease (CJD) was discovered in the early 1920's by two German Psychiatrists, Alfons Jakob and Hans Creutzfeldt.

It is one of four TSE diseases that occur in humans. The others are Gerstmann-Straussler-Scheinker syndrome, Kuru, and fatal familial insomnia.

Gerstmann-Straussler-Scheinker syndrome is an inherited disease that affects some 50 extended families. Loss of coordination precedes dementia and the course of the disease can take up to 6 years before death.(Warren Judd 1996)

Kuru occurred in the remote Fore natives of the New Guinea Highlands. In the Fore language kuru means 'shivering'. The spongiform brain changes and neurological and muscular symptoms are exhibited along with behavioral and personality changes. The disease was prevalent among women and children of the Fore tribes, attributed to the ritual cannibalistic practices that accompanied the care of the dead. It was common practice for the brain of the dead to be eaten as a rite of mourning and out of respect for the dead kinsmen. Women and children were the main mourners.

The incidence of Kuru has declined since the 1960's probably due to the decrease in the cannibalistic practice. (Merck Manual)

Fatal Familial Insomnia is another human disease akin to CJD. Untreatable insomnia progresses to overall impairment of the body's nervous and muscular systems. In advanced cases the patient may have hallucinations which is followed by coma and death. The mean age is 49 and the course of the disease takes about a year. (Warren Judd 1996)

Creutzfeldt-Jakob Disease is a progressive degenerative neurologic disorder, characterized by spongiform changes in the brain.

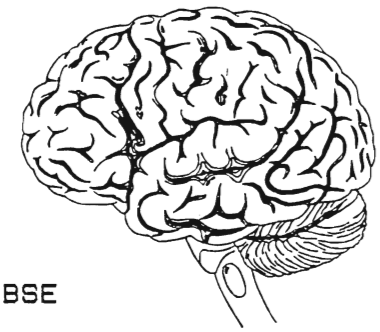
CJD occurs worldwide at a rate of between 0 - 5 per million people per year.

It is characterized by a late age onset of 60 - 79 years. The youngest person diagnosed was 17 years old. It has a long incubation period of many years. The disease ends in death after a 3 - 12 month illness. Some patients with clinical CJD remain ill for up to 2 years.

The disease is progressive, fatal and untreatable.

The signs and symptoms of the disease are confusion, progressive dementia, ataxia (failure of muscle co-ordination) and loss of reflexes and sphincter control.

2.4 INFECTIVE TISSUE



SCRAPIE	BSE	CJD
Brain Spinal cord	Brain Spinal Cord Eye	Brain tissue Spinal fluid
medium risk -		
Embryo Placenta Tonsil Spleen Lymph nodes	Ileum	cornea, kidney, lymph, pancreas, lung, liver, placenta
not detected or risk unknown		
Blood, heart muscle Kidney, mammary gland colostrum Muscle, milk, serum testis	Colon thymus bone marrow blood, heart Muscle, milk testis	Blood Urine faeces sweat saliva sputum milk

3.0 THE PRION THEORY

Among the scientific work and the media hype about BSE and CJD, one point is commonly agreed upon - that the exact cause the spongiform encephalopathies is unknown. No causal agent has been isolated or seen under microscopes.

The original idea was a slow acting virus but it soon transpired that the causal agent of the diseases lacks genetic material, that is, it does not have DNA or RNA in which to replicate itself.

Dr. Stanley B. Prusiner of the University of California evoked a great deal of skepticism in the early 1980's with this notion. He suggested that the diseases were caused by infectious agents made of protein and he named them Prions.

So what are Prions?

Brains of normal mammals contain a prion protein, PrPc, which is usually located on the cell surface of nervous tissue. This protein is coded for by a gene (PrP gene), found on chromosome 20 in humans. The function of PrPc is not known.

In the case of spongiform encephalopathies, the PrP gene is altered and is coded as PrPsc.

So if the infective agent that causes TSE's is a protein and does not have genetic material to establish infection in a host, then how is it spread?

Prusiner and his colleagues concluded that the Prions multiply in an incredible way. They convert normal protein molecules into dangerous ones simply by inducing the benign molecules to change their shape.(Prusiner 1995)

Prusiner's prion theory was radical to say the least. However, as time has gone by Dr Prusiner's explanation has been accepted.

Proteins are long, chain-like molecules, and the chain can fold up in different ways. What is different is that non-functional Prion proteins force functional prion- proteins to change shape and become non-functional.

A French scientist Dr Corrine Lasmezas and her team challenge Prusiner's theory. They have found that it is possible to transmit BSE to mice and then from one mouse to another without any sign of the unhealthy prion protein building up. The mice developed a full range of BSE symptoms but did not show any signs of the Prions. Given time, all the mice allowed to live long enough, developed the prion protein. Dr Lasmezas is suggesting that prions are not the cause of BSE but rather another symptom of it. Some other agent is doing the actual infecting.

The Economist Jan 1997

In October 1997 it was announced that Dr. Stanley Prusiner was awarded the Nobel Prize for Medicine for his work on the Prion theory.

3.1 DESTRUCTION OF THE INFECTIVE AGENT

Transmissible Spongiform Encephalopathies are fatal, untreatable and there is no apparent host immune response against the disease.

Scrapie has been found to be resistant to inactivation by heat, formaldehyde, ultra violet light, freezing or radiation(Bruere & West 1993)

CJD agent is resistant to formalin and standard sterilization (The Merck Manual 16th ed.p210)

The infective agent that causes BSE is resistant to standard sterilization methods also.

The agent can be inactivated by intensive prolonged periods of heat ie, Autoclaving 134oC for 15 minutes or by a cleaning solution of Sodium Hypochlorite.

3.2 DIAGNOSIS - IMMUNITY - TREATMENT ???

Currently work being done on a diagnostic test for BSE. At present diagnosis is made when an animal dies and the brain is dissected for analysis. The appearance or absence of spongiform holes confirming or negating the disease's presence.

The brain protein 14-3-3 is released from a CJD infected brain. The presence of this protein is detectable in the spinal fluid. However, as a diagnostic tool, this test is not solely conclusive.

There is no apparent immunity against TSE's.

A Japanese scientist, Shirou Mohri, removed the spleen and thymus (both important for immunity) from a mouse and then injected it with scrapie. It made no difference in the development of the disease, in other words no antibodies were made against the disease.(Dealler 1996)

For development of a TSE disease, there has to be a certain level of infectivity reached for the animal to exhibit symptoms - it is cumulative. Give an animal small amounts of infected material and when a certain level is reached then it will exhibit signs of the disease.

This may be the cause of great concern in the case of humans ingesting infected beef.

At present, there is no treatment for transmissible spongiform encephalopathies.

Experiments done in England by Christopher Budge and Mark Purdey, on cows with BSE, found that the symptoms of the disease temporarily relapsed with the use of nerve gas anti-dote.

4.0 TRANSMISSION OF SCRAPIE

According to Bruere & West (1993) the scrapie agent can be found in a range of sheep tissues. These include central nervous system tissue such as the brain, although it was not found until 25 months of age. Lymph nodes such as tonsils were found to have the scrapie agent also. However, the important findings from the Agriculture and Food Research Council's Neuropathogenesis Unit in Edinburgh, suggests that the scrapie agent may be transmitted via embryos. Their experiment consisted of transferring embryos from scrapie infected sheep. Five of the 20 lambs born developed scrapie. The scrapie agent was also found in full term placenta.

This means that the scrapie agent can be spread laterally to other ewes (see figure 1.1)

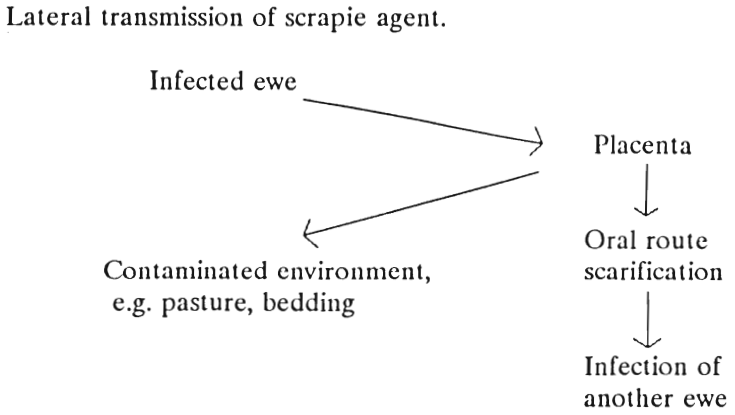


fig 1.1 Lateral transmission of scrapie agent (Bruere & West 1993 p359)

It can also be spread to the lamb pre and post natally. (see figure 1.2)

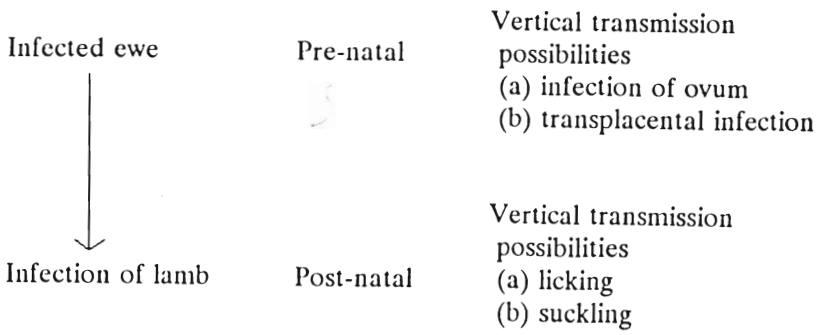


Fig 1.2 Maternal transmission of scrapie agent (Bruere & West 1993 p 360)

The ram does not appear to play a major role in the spread of the scrapie agent, according to Bruere & West(1993). However, scrapie was found in a Suffolk ram in 1952 in New Zealand. The ram had been imported from Britain in 1950 and in 1954 scrapie occurred on a Southland farm as a direct result of inadequate containment. It does not conclude whether the infection occurred via breeding from the ram or whether the ram and/or progeny had contaminated the farm environment. (Sheep Prod. vol 2)

Indirect contamination by the environment is possible, however, the method being via yards, needles, docking knives, feeding troughs and buildings.(Bruere & West 1993)

4.1 INCIDENCE OF SCRAPIE IN NEW ZEALAND

The United Kingdom would appear to be the main source of infection in the three isolated cases of scrapie in New Zealand.

Scrapie has existed in Britain for at least two hundred years and when we consider the fact that the majority of sheep flocks in NZ were derived from Britain in the last century, it is hard to believe that we are scrapie free.

It would appear that good fortune may have played a hand in the absence of scrapie in New Zealand. It may also be that the long sea voyage of up to six months may have acted as an enforced quarantine and infected ewes would have been disposed of at sea. In the 1850's large numbers of Merinos were sourced from drought striken areas in Australia and they were considered to be scrapie free.(Bruere & West 1993)

The Suffolk sheep would appear to be a breed with a high incidence of scrapie in Britain so why did it not appear in NZ - considering they have been imported in to NZ since 1913 ? Small flock numbers may account for this as is the probability that the sheep imported were luckily scrapie free.

As stated earlier, a Suffolk ram brought in to NZ in 1950 developed scrapie in 1952. Scrapie then appeared on a Southland farm in 1954 as a result of inadequate quarantine and follow-up.

> ' A serious containment exercise ensued which lasted for three years until eradication was declared. It involved the destruction of 4339 animals and the listing and movement control of stock from 191 farms for three years.'
>
> Bruere & West 1993:364

Scrapie was again diagnosed in 1976 and 1977. The two cases were from a Finn sheep and an East Friesian sheep, both of which were in quarantine on Mana Island. They were part of a breeding exercise in exotic sheep breeds which was subsequently curtailed and the events which followed saw the destruction of several thousand sheep on Mana Island and Crater Farm in Rotorua.

4.2 SCRAPIE IMPORTATION

The risks of further out breaks of scrapie in New Zealand are considered to be minimal due to strict quarantine and regulations surrounding the importation of sheep.

In early 1997, AgResearch New Zealand applied to import scrapie infected sheep brain into the country. The reason given for this was that more modern techniques of testing, such as western blot, could be used to convince and persuade European governments that New Zealand is scrapie free. However, at present New Zealand does not have the facilities to contain the disease. The application was denied by the Agricultural Security Consultative Group who were given the job as judge and

jury by the Ministry of Agriculture. Apparently the European Union is refusing to acknowledge that NZ is scrapie free because they argue that NZ does not use adequate methods of detection of scrapie or BSE.

Alternative solutions are being looked into by MAF's Chief Veterinary Officer, Dr Barry O'Neill.

Scrapie has been a notifiable disease in New Zealand since 1955.

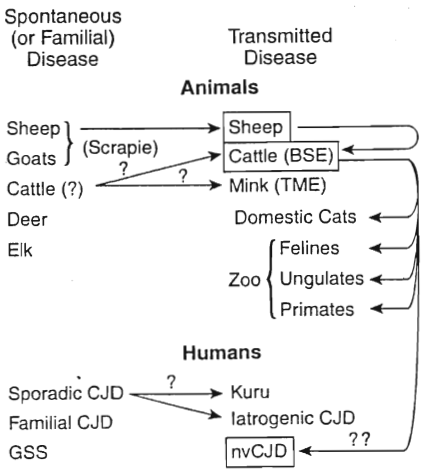
5. TRANSMISSION OF BSE

Experimentation has shown that the brain, the upper spinal cord, and the eye are the only parts of cattle with BSE which could infect another animal.

The experiments could not detect infectivity in any of 45 other bovine tissues, including muscle (meat), peripheral nerve tissue, intestine, lymph nodes, testis, prostate, semen, ovary, udder, milk, uterine caruncle, placenta or placental fluids

(Royal Society of New Zealand June 1996)

BSE infective tissue has been transmitted to other mammals. Cats, mice, kudu are 3 of 14 animals that have become infected from BSE tissue. (fig 1.3)



Brown (1997) JAMA 278 no.12 p1029

A study done by John Collinge et al at St Mary's Hospital, London, has looked at the PrP make up of the various animals that have contracted BSE infection , and at the common CJD and new variant CJD. They have discovered that new-variant CJD has strain characteristics distinct from other types of CJD and which resemble those of BSE transmitted to mice, domestic cat and macaque, consistent with BSE being the source of this new disease. Further trials and studies are being done to confirm these findings as are the possibility that BSE may have been transmitted to and is surviving in the sheep population

(Nature vol 383 24 October 1996)

The other aspect in the CJD and BSE scene is the lack of simple diagnostic techniques.

An experiment in Yorkshire, UK was set up by MAFF to determine whether BSE could be vertically transmitted either from sire and/or dam to their progeny.

New Zealand was chosen to provide 350 Friesian X Hereford heifers because of the absence of Transmissible spongiform

encephalopathies associated with livestock. The maiden heifers were imported into Britain at 9 months of age in 1990 and 1991 to be recipient dams for embryos collected from BSE infected cows.

The New Zealand heifers were implanted with embryos at 22 months of age. From these embryos there are 120 breeding females. As yet none of the animals are old enough to display BSE symptoms. The experiment is expected to conclude in the year 2000. (Rural News April 1 1996)

Claire Powell, the UK reporter for the Rural News reports that on August 1, 1996, the UK MAFF announced that interim results from a trial had shown that maternal transmission of BSE may be possible.

The trial was carried out by the Central Veterinary Laboratory at Weybridge in the United Kingdom. It involved two groups of more than 300 animals.

Group A consisted of offspring of confirmed cases of BSE. Group B comprised offspring of cows from the same herd who had not developed signs of BSE at the age of at least six years. The animals from both groups were kept until age seven or until BSE or another disease intervened.

BSE occurred in both groups.

The following are the results of the trial

	GROUP A	GROUP B
Offspring of cows with BSE	+	-
No. of cattle	300	300
No. of cattle to reach 7 years	273	273
No. of cattle to develop BSE	42	13

The Spongiform Encephalopathy Advisory Committee (SEAC) reviewed the report into the study of maternal transmission of BSE.

It concluded that the evidence is not conclusive by any means but the possibility does exist that BSE can be transmitted to offspring.

Just how the disease is transmitted from cows to calves was not indicated in the trial.

Scrapie in sheep can be passed on from dam to progeny via the placenta.

MEAT AND BONE MEAL

Meat and Bone meal is the powdered remains of animals, and is used as either fertilizer or stock feed.

It usually consists of waste material or by-products, after all usable material has been removed from an animal.

FEED BAN

Since June 1988, all possible cases of BSE had to be notified to UK MAFF. Since July 1988, the feeding of ruminant protein to ruminants has been prohibited. Soon after this a ban, known as the Specified Bovine Offal (SBO) ban was put in place. It relates to specific tissue such as brain, spinal cord, tonsils, spleen and thymus. Muscle tissue, milk, and gelatin products have not been shown to transmit the BSE agent.

(Public Health Report Apr 1996:26)

The SBO ban was in response to UK MAFF's acceptance that the meat and bone meal was spreading the BSE epidemic. They calculated that the incidence of BSE should start to decline from 1993.

5.1 IMPORTATION OF BOVINE PRODUCTS

The question about whether it is safe to import bovine embryos and semen from the UK has been asked many times.

An independent scientific advisory panel was set up to look at the issues surrounding importation of cattle embryo's and semen. Mr Barry O'Neill, New Zealand MAF's Chief Veterinary Officer, stated that MAF believes it is safe to import bovine genetics from the UK. However, a suspension of such imports has been put in place in response to what he calls 'hysteria' linking 10 UK cases of CJD and BSE.

Initially, bovine embryo's and semen were banned from NZ in 1989 following the outbreak of BSE .

(Live cattle imports were banned in 1988)

The ban was lifted in 1992, the same year that standards were imposed in relation to importing bovine genetic material. These set of standards were devised by the World Animal Health Organisation

6.0 ORGANOPHOSPHATE LINK WITH BSE - conspiracy or coincidence ?

Claire Powell, the UK reporter for Rural News regularly updates New Zealand readers with the latest news on BSE.

In July 1996 she writes of the Mark Purdey's organophosphate theory.

> "For a number of years Mark Purdey, a farmer and scientist from south West England, has publicly and vigorously maintained that chemicals compulsorily used in the 1980s and early 1990s to eradicate warble flies, have played a significant role in the BSE situation."

The warble fly is an external parasite that lives on cattle.

The chemical used in the warble fly dressing was a pesticide called Phosmet. It was a combination of organophosphate and phthalimide (also used in thalidomide the drug prescribed in the 1960s to treat the symptoms of morning sickness in pregnant women. The resulting side effects of thalidomide on the foetus caused birth defects - commonly deformed or missing limbs)

The warble dressing (as it is come to be known) was poured along the backs of cattle to poison the warble larvae by attacking its central nervous system. MAFF regulations stated that the warble dressing had to be applied between March 15 and July 31 of every year. The use of phosmet declined in the early 1990s as the warble fly was eradicated. Mark Purdey believes that the chemical dressing was responsible for the BSE epidemic. He believes that the cause of the problem is that the chemical gets into the brain and binds with normal prions to produce abnormal ones.

Purdey also believes that the chemical can attack an unborn calf which may account for why some calves have been born with BSE although their mothers did not succumb to the disease.

He does subscribe to the theory that the Prion is the infectious agent in BSE but argues against the theory that meat and bone meal caused the outbreak of BSE. Purdey believes that it was the use of organophosphates that have caused the abnormal prion changes.

This may account for the fact that although all cattle in a herd ate the same feed - not all got BSE and since the ban on contaminated feed in the UK, there have been approximately 30,000 confirmed cases of BSE in cattle born after the ban. These cattle may have been damaged as embryos by the warble dressing.

The warble fly treatment with Phosmet was a compulsory practice imposed by UK MAFF. Phosmet was applied at a dosage of four times the manufacturers recommended safe dosage. This went on for approximately ten years - from 1980 to 1990.

Purdey's attempts to convince the British Government of his theories have proven to be somewhat futile. It would appear that British MAFF are not about to accept the OP theory (Rural News Dec 1996)

It would not look good for MAFF if their compulsory, high dosage campaign against the warble fly was proved to have caused this massive epidemic of BSE. The legal claims and litigation suits would keep lawyers employed for ever and a day.

So, was the infected meat and bone meal theory a smoke screen of protectionism?

Is the OP theory the cause everyone has been looking for?

This was not the first time Mark Purdey had had a run in with MAFF. Purdey and his lawyer Peter Ward defeated MAFF in 1984 when MAFF tried to force Purdey to use Phosmet on his organic cow herd (subsequently none of the herd developed BSE)

Peter Ward was an environmental legal lawyer who was subsequently killed in a mysterious car accident.

Purdey's vet Christopher Budge who assisted him in trials of nerve gas antidote in treating BSE cows, died in a head on car collision.

Mark Purdey himself, had his barn burnt down, shots fired at his house, his phone lines cut, and his house burnt down.

Dr C. Bruton, a scientist associated with prion research and discoverer of the new variant CJD strain was also killed in a vehicle accident.

A Japanese scientist gunned down in the USA had copies of Purdey's theories.

It would appear that subscribing to controversial theories might be hazardous to your health!.

Apparently there have been no cases of BSE in organic beef herds. Purdey's organic herd were fed meat and bone meal but were not treated with Phosmet and he has no cases of BSE.

It has been claimed that the BSE epidemic was a scandalous cover up to protect the pesticide chemical companies and the UK Government, and is believed to be substantiated by the following facts.

1. BSE levels are particularly high in areas where high OP pesticide applications are mandatory on cattle in warble fly control areas in the UK
2. No BSE has occurred in organic dairy herds where stock have been drawn from an organic pool.

Soil & Health 1996

Two other countries who have license to use Phosmet are Ireland and Switzerland. These countries are second and third to the UK in the BSE league.

An argument against the OP theory is that as the warble fly dressing was compulsory then why haven't as many beef cows as dairy cows been affected? Supporters of the Purdey theory maintain that the extra fat levels of a beef cow enable them to absorb more OP in their fat, acting as a buffer against the full impact of treatment.

Some scientist will go part way in their support of the Purdey theory by saying that the warble dressing may have reduced the resistance of the treated animals to the infective agent which causes BSE (Claire Powell July 1996)

Phosmet was licensed in New Zealand for orchard use by Turners and Growers until 1991, however, registration has since been cancelled.

Mark Purdey also believes that people who have been exposed to high levels of OP may be at risk to the likes of CJD. He also believes that the likelihood of a beef eater developing the human version of BSE is remote, unless their own immune system has been compromised by contact with an OP such as Phosmet.

One would think that the UK Government could use the theory to take some of the heat off the panic stricken beef markets. It would be an alternative to infected beef causing the new variant CJD.

However, Mark Purdey's repeated attempts to obtain research information and to communicate his beliefs with MAFF have been futile or thwarted. The UK Government simply will not buy the OP theory, for whatever reasons.

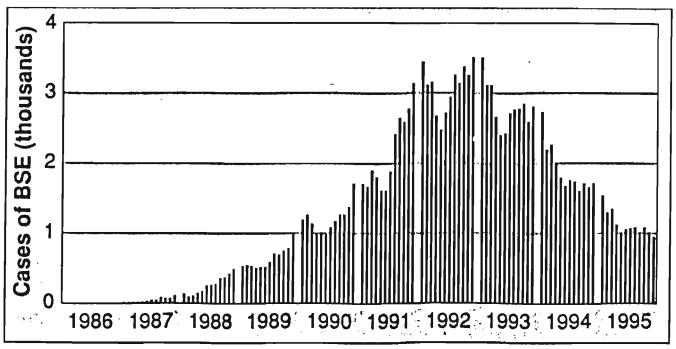
7. BSE EPIDEMIC

7.1 EFFECTS ON UK

The BSE epidemic has been described as the biggest epidemic of disease that Britain had ever seen.

Features of the epidemic are listed below:

- it has predominantly affected dairy breeds and the majority of cases are in the south of England.
- Cows are more likely to have the disease than bulls - 90% cows, 10% bulls.
- also know as 'Mad Cow Disease' because of the strange behaviour exhibited by affected cows. A cow may become very nervous, lick their noses and grind their teeth a lot. They also hold their heads down, arch their backs and sway and trot instead of walking normally, and lift each leg up much higher than usual. (Science behind the News)



The epidemic. Confirmed UK cases of BSE in cattle by month of onset.

It was calculated that around 350,000 head of cattle would develop the disease up until the year 2000.

1st case of BSE confirmed in November 1986, on a farm in the south of England.

By 1987 the disease had spread and it looked like it was not going to go away

On 21 June 1988 BSE became a notifiable disease to UK MAFF

Ruminant derived feed ban was imposed in July 1988.

Since 1986, more than 167,000 cattle have died of BSE in Britain, with over 30,000 BSE cases in animals born since the ruminant protein ban was implemented in 1988.

The latter is probably due to either leakage in the ruminant feed ban and the SBO ban or maternal transmission being the cause.

A further ban was imposed by the European Union as a result of pressure from Germany. No beef from animals over 30 months old was to be sold for human consumption. The EU banned the export of beef, beef products and live animals to other EU states and third countries. (It had been found that British beef has since been sold illegally to certain third world countries)

Since the ban on 30 month and older cattle for consumption, over 1.2 million cattle have been slaughtered (Agra Europe Feb 1997)

7.2 UK GOVERNMENTS RESPONSE

COMPENSATION

The British Government introduced compensation on 8 August 1988 for farmers with cattle considered to suffer from BSE. This was set at 50% of the value of the cow.

By the end of that year, 1988, 527,142 pounds had been paid to farmers in compensation for BSE.

MAFF announced compensation figures in February 1990. The average market value of a milking Friesian cow and calf was 645.82 pounds. If a cow was found to have BSE then 645 pounds was paid out. If by post-mortem BSE was not found then 818 pounds was paid to the farmer. The final price was agreed on between a Veterinary Officer and the farmer.

2,826,788 pounds was paid out in compensation in 1989

By the end of February 1990 4,135,729 had been paid to farmers in compensation of BSE. Also by this time, 5.1% of all herds had been affected by BSE.

By the end of 1990 9,114,743 pounds was paid out in BSE compensation. (Dealler 1996)

EU imposes a world wide ban on all British beef imports March 1996 after the announcement of the new variant-CJD

The ban also extended to 'at-risk' materials from sheep, cattle , goats and cosmetics - world wide. This affected New Zealand's exports of tallow, a by-product of meat processing.

While NZ is BSE and scrapie free, the European Commission does not acknowledge this fact. However, Tallow has been given the all clear as far as transmission of TSE's go.

THE POLITICS

Many scientists and researchers who were keen to find answers to the bse epidemic were thwarted in their efforts. Many, such as, Dealler and his colleague Lacey, had serious misgivings about the continual reassurances from the UK Government that eating British beef was safe. However, their efforts to convey their feelings and theories were ignored. Instead of money being poured in to research and experimentation, money was spent on massive campaigns of unfounded assurances. The scientists argued that it could not be proven that beef was safe to eat during the epidemic and the Politicians argued that it had not been proven that it was not safe. Dealler's book 'Lethal Legacy' outlines in detail his efforts to pursue concerns regarding BSE and efforts thwarted or down played by the Government, UK MAFF and the Meat and Livestock Commission.

In March 1997, Colin Maclean, the Director General of Great Britain's Meat and Livestock Commission, attended the New Zealand Meat Producers' Board annual general meeting. He addressed the conference and made reference to his country's BSE epidemic. According to Mr Colin Maclean food safety was on the top of consumers' needs list, ahead of welfare or environment issues. After a political nightmare regarding eggs and salmonella, listeria and pate, and e. coli Britain was very sensitive to food scares and initially this would seem to be the case. Soon after the announcement that a new variant strain of CJD had been isolated with its possible links to BSE, the beef market plummeted.

Cattle sales through markets went from about 22,000 a week before the announcement, to only 700 in the week after the announcement. Beef consumption fell almost 60%. Mr Maclean believes that faith in the industry was restored by introduction of a Quality Minced Beef Mark and reassurances that eating British beef was quite safe. Interesting to note that mince accounted for almost 40% of total beef sales. Although beef consumption has gained ground over the last 12 - 18 months, sales are down by around 15 - 20 % of what they were before the March 1996 announcement that BSE and new variant CJD may be linked.

The MLC's objectives are to rebuild consumer confidence, improve quality and communication and maintain price competitiveness. Was this desire to rebuild consumer confidence based on the need to maintain a market or to provide the consumer with a safe product?. Mr Maclean states that "by fully understanding consumers attitudes and designing programmes that directly address their needs and aspirations of the product, then we can influence their buying decisions." (Maclean 1997)

However, as time went on, food safety became less of an issue, and the price of beef became important.

Interesting to note that as the price of beef in the UK dropped, butchers and restaurateurs reported the public returning to eating British beef.

Another difficulty that researchers had was actually obtaining samples of infective brain tissue. THe Ministry of Agriculture, Fisheries and Food (MAFF) own all the bodies of cows infected with BSE and controls all the data relating to the disease. For scientists outside MAFF they have had great difficulty obtaining samples or data.

Two scientists in California have what they believe a simple diagnostic test for BSE. The test works by identifying a protein which leaks in to the spinal fluid as a result of brain damage due to either BSE or CJD. However, they have had only 10 samples on which to base their work. No further samples from MAFF were forthcoming. At present the only way to diagnose BSE is by post mortem examination of the bovine brain - an expensive and possibly risky procedure (20 - 30% of brain samples test negative to BSE)

" It would appear that the BSE crisis is a case study on how not to handle an epidemic. Politics seems to have got in the way of science." (The Economist Feb 22 1997 p67)

In February 1997 'The Times' also reported a test that had been developed by a diagnostic company in Dublin. The test also detects the presence of Prion protein in spinal fluid.

Both of these test may detect the presence of BSE, however, the disease would by then be established in the host.

The British government's concern was with the blanket EU ban on British beef and beef products. In response to this in May 1996 British Parliament rushed through measures to try and restore consumer confidence and to reduce the future numbers of cattle developing BSE.

The regulations state that all carcasses and meat from all animals slaughtered at more than 30 months of age are to be excluded from all human and food chains. It is anticipated that the ban will remove around five and a half million cattle from the British beef industry over the next six years.

Since the introduction of these measures, the British Agricultural Minister, Douglas Hogg, went in to bat against the European Parliament with a further proposal to kill all cattle born after 1990 from herds that had had BSE. What he came back with was yet another set of conditions from the EU. Cattle targeted in this slaughter scheme were called cohorts in that they were calves raised with other calves that have since developed BSE.

The realities of putting such schemes in to place has been a nightmare for farmers, abbattoirs, vets and the like.

Determining the exact age of some cattle is not easy. Organic herds who are over 30 months of age but have never had BSE in them are slaughtered as unfit for human consumption as are late finishing, grass fed herds.

In effect, this meant that a 29 month old pure Holstein or Ayrshire steer was eligible for human consumption, whereas a 30

month and one day old Highland steer, reared on heather and grass on a Scottish Island was not.

A further problem appeared in that there appeared to be a lack of organisation in relation to the killing of 30 month plus cattle. Incineration is needed to dispose of the carcasses and these facilities were not available to handle the proposed 30,000 head of cattle a week. The abbattoirs were not informed of their role and in any case their freezers were all ready full of beef which nobody wanted.

A 'bottle-neck' situation had arisen with in the industry and the flow on effects to the community were being felt. The situation was affecting abattoirs, livestock transport companies, auction marts, feed companies and many small rural businesses.

However, by July 1996, 200,000 head of cattle had been disposed of with another 200,000 waiting their turn.

By January 20th 1997, 1.18 million cattle had been slaughtered under the UK Government's over-30-month scheme.

Compensation for the 30-month and older scheme is set at market value while the UK Farmers Union had been calling for compensation to be set at replacement value.

The number of BSE cases confirmed in the UK between 1988 and the end of 1996 totalled 167,089.

A German Member of Parliament commented wryly that

" the British politicians are keener to fight the European ban than to fight the real problem - BSE"

If we were to ask many of the scientists and researchers, they would probably agree. In many cases, funding has been withdrawn from research and barrier's put up to prevent scientists communicating their findings and concerns. Dr. Stephen Dealler, a consultant Microbiologist, went so far as to write a book on his frustration at the UK Government for their attitude to scientific research on the subject of BSE.

7.3 EFFECTS ON NEW ZEALAND AND RESPONSE TO BSE

In August 1996, in response to the ban on British beef and beef products, Barry O'Neil, Chief Veterinary Officer of the Ministry of Agriculture, issued a statement relating to New Zealand's status regarding scrapie and BSE.

He states that NZ is and always has been free of BSE. It is also free of Scrapie.

Transmissible spongiform encephalopathies have been notifiable diseases under the Biosecurity Act 1993 - scrapie has been notifiable since 1955 and BSE since 1989.

- Importation of any livestock food containing meat and bone meal is prohibited.
- Border control at all ports of entry into the country to monitor the importation of all animals, plants and their

products.

- In response to the BSE epidemic importation of live cattle, bovine embryos and semen were suspended in December 1988, then again in May 1989 and all but live cattle imports resumed in October 1993

Suspensions were again put in place in March 1996 following possible link with BSE and CJD.

Barry O'Neil's objective with this document was to convince the European Union that NZ had strict policies regarding bovine imports and that NZ is free from scrapie and BSE.

However, the European Commission has not acknowledged this and continues to state that NZ can not prove that we do not have BSE or scrapie. (Hence the attempt to bring scrapie in to NZ in 1997 in order to prove that we do not have it.

WHAT EFFECT DID THE BSE EPIDEMIC HAVE ON NZ EXPORTS?

The London office of the New Zealand Meat Board have reported increased sales of New Zealand lamb since the March 20 1996 announcement that BSE and a new variant form of CJD may be linked. UK lamb retail sales rose by around 13 per cent where as NZ lamb sales rose by 43 per cent. (Rural News May 1996)

8. CREUTZFELDT-JAKOB DISEASE (CJD) - A HUMAN TSE

8.1 FORMS OF CLASSICAL CJD

INHERITED CJD

This form of human spongiform encephalopathy is due to mutations of the PrP gene found on chromosome 20.

IOTROGENIC CJD

Iotrogenic means brought about by surgical/medical treatment for example, unwanted effects of drugs.

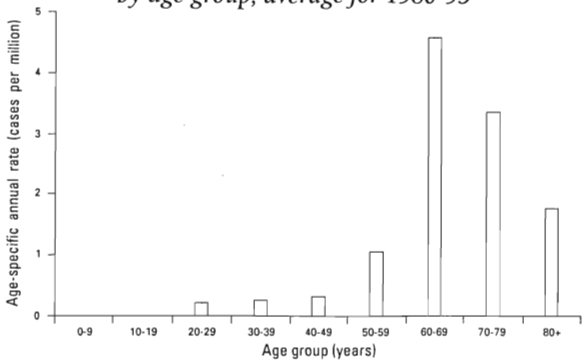
This form of CJD has been brought about by direct infection via tissue, hormone transplants and implants.

Replacement therapy of Growth Hormone (GH) was given to children of short stature and those with a GH deficiency. The semi-purified GH was prepared from human pituitary glands. However, the purification procedures did not inactivate the agent which causes the TSE and CJD was transmitted to these patients. This human derived hormone is now banned and a synthetic growth hormone has replaced its use. Human to human transmission also occurred during corneal and dura mater transplantation and by use of contaminated brain electrodes. (Merck Manual 16th ed. p209-211)

SPORADIC CJD

There is no mutation of the PrP gene with this the most common form of CJD. In the majority of all cases of CJD , the reason for the disease remains unknown.

Creutzfeldt-Jakob disease rates by age group, average for 1980-95



As previously mentioned, the preceding forms of Creutzfeldt Jakob disease usually affect people in the age range of around 60 to 79 years of age.

8.2 INCIDENCE AND SURVEILLANCE OF CREUTZFELDT-JAKOB DISEASE

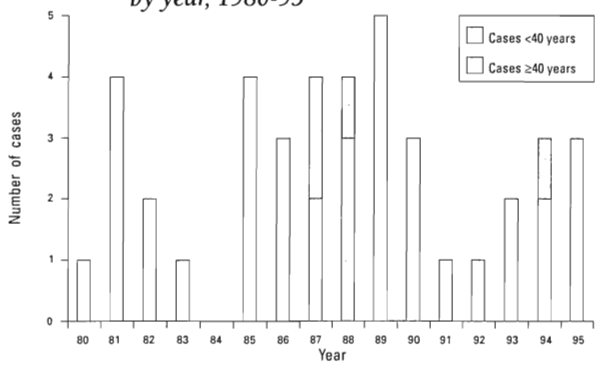
In New Zealand the incidence of CJD averages out at 0.8 cases per million people per year (between 0 - 5 per year)

There were 41 cases of CJD between 1980 and 1995. 23 females, 18 males and 95% were non- Maori. (Weinstein et al 1996)

Of the 41 cases diagnoses of CJD some were made on clinical history alone - not confirmed with histopathological investigations. That is, post mortem tissue sampling of the Central nervous system was not carried out.

When looking at the signs and symptoms of CJD it could easily be confused with Dementia or Alzheimer's Disease, however, the short duration of the illness and a characteristic EEG (electroencephalogram) set it apart.

Creutzfeldt-Jakob disease cases by year, 1980-95



17% of the 41 cases in New Zealand were linked to growth hormone treatment, 10 - 15% were inherited and the remainder are not linked to any known factor (sporadic).

Infected tissues with CJD are the brain tissue including pituitary hormones, dura mater, and cerebral spinal fluid.

Brains of normal mammals contain the prion protein, PrPc which is found on the cell surface. This protein is coded for by a gene (PrP gene) found on chromosome 20 in humans. (Infection control policy 1996)

8.3 NEW VARIANT CJD

In March 1996, it was announced by the British Ministry of Health that there had been a new development in the CJD disease. The CJD Surveillance Unit in Edinburgh informed the public that ten cases of a different type of CJD had appeared. The alarming factor of these 10 new cases was the relative youthfulness of the patients. They were aged between 19 and 41 with a mean age of 29.

While the symptoms exhibited were similar to normal CJD, there were some differences. For example, the typical EEG appearance is not there, the disease being more prolonged and patients present with psychiatric symptoms.

Of these 10 cases of nv-CJD, four male and six female, only two remained alive by March 1996. Information on nine of the cases is as follows. Four had no history of any operation and the other four had various minor and major operations. None were from farming backgrounds and all had eaten beef or beef products in the last 10 years. One person had been a strict vegetarian since 1991. (Lancet 1996; 347:921 25)

To date there have been no cases of new-variant CJD reported in New Zealand.

The most likely explanation for the new-variant CJD has been exposure from the BSE agent. The possibility exists that transmission of the BSE agent occurs by ingesting tissue such as brain, spinal cord, tonsils, spleen or thymus infected tissue. This has not been proved but the UK Spongiform Encephalopathy Advisory Committee (SEAC) announced that the possible link was " cause for great concern".

Subsequently, the youthfulness of the patients and the presence of many large 'daisy' shaped plaques in the brain also pointed to BSE being the causal agent of this new variant strain of CJD. Normal CJD rarely shows wide-spread plaques(Manuelidis et al 1997)

Up until 1996 the UK Government had consistently been giving unfounded reassurances to the public that eating British beef was safe.

Then following the discovery of the nv-CJD in March 1996 they had to acknowledge that they could not prove that eating British beef was safe from BSE.

The CJD Surveillance Unit was set up in Edinburgh in 1990 to closely monitor cases of CJD.

By October 1996 a fourteenth victim of nv-CJD died in England. The latest information from the Edinburgh Surveillance Unit as at 3rd October 1997 has confirmed 20 cases of new variant CJD and

one probable case.

The World Health Organisation has also taken steps to attempt to limit the transmission of BSE and to reduce the possible risks to humans. A consultation group was set up to review the information available regarding BSE and the new variant CJD. They came up with recommendations relating to BSE and nvCJD. They reiterate many of the practices all ready in place. For example: banning of ruminant tissues in ruminant feed; no BSE tissue to enter the human food chain; milk is considered safe as is tallow and gelatin if rendering procedures are in place. More research is needed in to CJD and further data is required in to the possible link with nvCJD and BSE and also to increase worldwide surveillance for BSE and CJD.

In New Zealand a CJD surveillance group has been set up in Dunedin contracted by the Ministry of Health - July 1997.

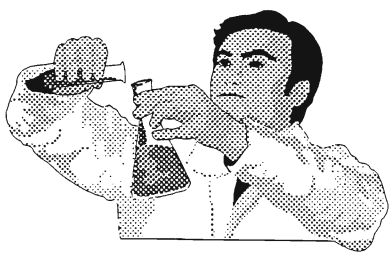
Led by Associate Professor Martin Pollock, their role is to keep watch on the incidence of CJD in New Zealand. They are notified of new cases of CJD and follow up is then made to ascertain whether the patient has ever donated blood. They also receive post-mortem reports in relation to CJD cases.

8.4 DIAGNOSTIC TESTS

In the area of Transmissible Spongiform Encephalopathies, the lack of specific and accurate diagnostic tests makes CJD difficult to diagnose ante-mortem.

A test does exist that can be definitive if other symptoms of CJD are present. Epstein and Brown (1997) report on the test where spinal fluid is analysed for the 14-3-3 brain protein released by the TSE-infected brain. However, the protein is also released when a person suffers a stroke so the presence of the protein is not limited to CJD alone.

In March 1997 the World Health Organisation updated their recommendations in relation to medical products. Medical products from bovine origin should be avoided or if this is not possible then source the bovine product from a BSE-free country that has an effective surveillance system. Medical products from human origin are considered safe as in the form of blood products except in the case of donors that have received extracts of human glands, brain matter transplants or have a family history of CJD.



8.5 COMPARISON OF CJD AND NEW VARIANT CJD

	CJD	new variant CJD
age of onset	60 - 79	18 - 41
duration of illness	3 - 12 months	7 - 24 months
incubation	up to 30 years	5 - 10 years
presenting symptoms	progressive dementia confusion	Psychiatric e.g. behaviour mood changes
	specific EEG changes	EEG changes not evident
leading to neurological involvement	ataxia	ataxia
	spongiform changes in the brain	spongiform changes in the brain
		PrP plaque formation (similar to BSE plaques)

There are many questions that arise in the discussion of BSE and the links to new variant CJD

One conundrum is that humans have been eating scrapie infected sheep meat for decades with no evidence that scrapie has caused CJD.

Scrapie and BSE are essentially the same disease in two different animal species so why should BSE contaminated products be infectious for humans when scrapie contaminated products are not?

A possible answer to this is that the nature of the infective agent has changed in the course of passing from one species to another (Kuru can be transmitted to ferrets only through non-human primates, and not when inoculated directly)Brown 1997

Therefore, scrapie may be infective to humans only after passage through cows.

Another puzzling observation is that the two most widely consumed products, meat and milk, do not transmit the infective agent in an animal with a TSE. So, how have humans supposedly acquired BSE?

Two possible answers exist. One is that the milk and meat may harbour the infective agent but it is not detected in the method used for testing for it. The second reason could be that meat is contaminated during slaughter. The saws and cutting instruments used for halving a carcass could disperse contaminated spinal cord tissue. The mixing of muscle with spinal cord to make meat products could have contributed to the spread of BSE to humans.

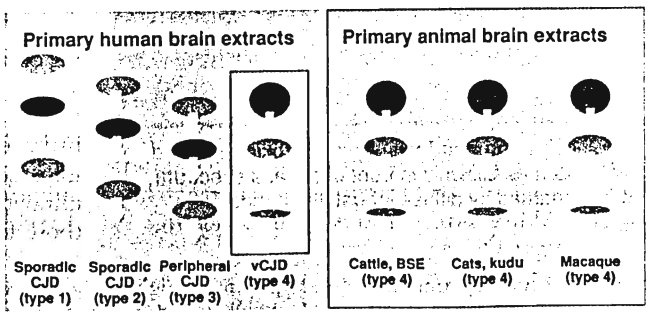
The third question that arises in this debate is why does the nv-CJD seem to target only young people when all age groups will have consumed beef products?

This question is the most difficult to answer. It is possible that for some reason young people are more susceptible to this infection or that they have a shorter incubation period that older people. Not conclusive by any means however, it is possible that more older people will be affected in the future.

It is also possible that poultry and pigs may be assisting the spread of BSE to humans but this has not been proved - probably because these animals are slaughtered at too young an age for the diseases to be evident. (Brown 1997)

It would also appear that TSE's only occur in mammals (mammal brains contain the normal prion protein) so poultry may never be indicted for the spread of BSE to humans.

Collinge and co-workers (1996) have performed extensive tests concerning the nature of the new variant CJD in comparison to normal CJD and BSE. They have consistently found, using Western blot preparations, that the pattern of new variant CJD is identical to BSE and quite different to normal CJD.



Spot check. Collinge's test shows clearly that the pattern of prion masses in new variant CJD mirrors that of BSE.

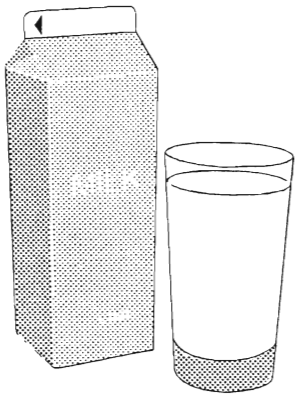
The hypothesis is that BSE and the new variant CJD are linked - the BSE agent is possibly the cause of the new variant CJD.

While there is still no conclusive link between BSE and nv-CJD, there remains cause for great concern and it is predicted that further cases of nv-CJD are likely to arise.

The period of time when BSE would have been most likely transmitted to humans, were the years preceding the Specified Bovine Offal ban and from when the rendering procedures changed in the late 1970's to early 1980's.

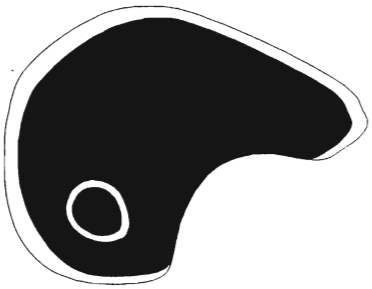
FOOD SAFETY

So, is it safe for the Brits to consume British beef and to drink cow's milk?

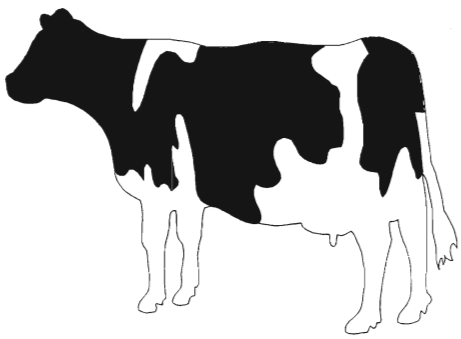


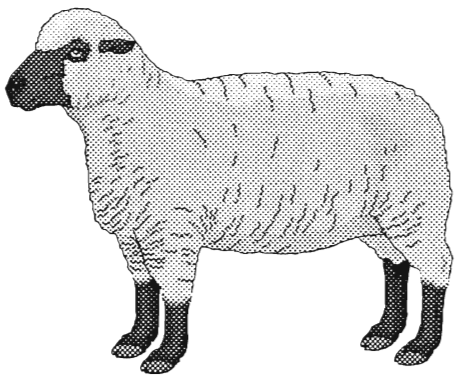
As meat and milk have consistently been demonstrated not to transmit the infective agent, then it is likely to be quite safe to consume. Taking into account that strict measures need to be continually enforced to ensure that infective tissues do not enter the production of consumer goods.

Epstein and Brown (1997) note, however, that transmission of prion disease by the oral route has been accomplished only after extremely high doses of inoculation. Ingesting the infectious agent is 100,000 times less efficient than directly into the brain of a host.



There are various beliefs held surrounding the possibility of new variant CJD becoming a raging epidemic. Dealler (1996) would suggest that it is quite possible that BSE was transmitted to humans and has been trying to get this message across for a number of years. Others too, such as Brown and Collinge, would also suggest that it is possible that BSE has being transmitted to humans. However, the lack of conclusive evidence means that there is still room for optimism that nvCJD may remain a very rare disease. Time will tell.





10. RECOMMENDATIONS AND CONCLUSIONS

There remain many unanswered questions about TSE diseases. What is known is: TSE's are progressive, they attack the central nervous system of the host, they are probably caused by an infectious prion agent and they are invariably fatal.

Research and experiments are still being carried out regarding transmission, method of infectivity, and expected duration and cessation of the BSE epidemic.

It is difficult to predict the scope of the incidence of new variant CJD without knowing the cause. As more is discovered and questions answered, then we may be able to better understand the nature of this merciless disease.

Further research is needed to ascertain the nature of transmission of the infective agent. To learn more about the diseases it is important to ascertain the nature of transmission for sheep, cattle and humans. There does appear to be host species barriers to the transmission of prion disorders. A possibility exists that humans may be being infected with scrapie but that the scrapie agent has been transmitted to humans through cows. Kuru has been transmitted to chimpanzees, monkeys, mink, ferrets and goats but not to sheep. Scrapie is transmissible to hamsters but not to chimps. It has also been suggested that humans have a specific barrier to BSE but these tests have not been substantiated.(Epstein & Brown 1997)

It is important New Zealand try to satisfy the European Union that New Zealand is BSE and Scrapie free. Maintaining strict surveillance and documenting of same will be helpful as well as continuing the ban on bovine embryos and semen.

Continuation of the protein ruminant feed ban is important in the UK and here in New Zealand.

Continuation of the ban on British beef products in to New Zealand must be upheld.

Surveillance of CJD in New Zealand must proceed and accurate information kept on victims of the disease. At present it must be assumed that blood may be able to transmit the disease, so CJD patients and families should not be able to donate blood.

Measures put in place regarding worldwide surveillance are important to learn more about the nature of new variant CJD. Funding, samples and access to information should be guaranteed to be available to those researching BSE and CJD.

Information and confirmation relating to the causes and nature of Transmissible Spongiform Encephalopathies is needed soon to understand this group of diseases that shows no mercy to its hosts.

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