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Tomorrow the world; Have contaminated feed experts spread BSE across the globe?; 10 February 2001

Germany utilises BSE crisis to implement EU plans to restructure agriculture; 31 January 2001

BSE/"mad cow disease" crisis spreads throughout Europe; 23 January 2001

Humans may get different forms of BSE;11 November 2004

Humans are likely to catch more than one form of mad cow disease, experiments in mice suggest. But the good news is that some people may have a genetic make-up that protects them against the disease.

So far, all 146 patients in the UK who have died of the human form of mad cow disease had a distinctive type called variant Creutzfeldt-Jakob Disease, or vCJD. **And they all have a similar genetic background for the "prion" protein implicated in the disease - called an "MM" genotype. This is present in 36% of the UK population.**

The results from the mice experiments now suggest that at least one, and possibly two, other forms of the disease might exist in people. **And the new forms may affect people with the remaining two genetic backgrounds: MV, which is in half the population; and VV, in about 14% of the population**. The new research is published in *Science*.

"It means that the human population won't have just one form of disease associated with BSE infection," says Jonathan Wadsworth of the UK Medical Research Council's Prion Unit in London, who is an author of the paper with team leader John Collinge.

Abnormal proteins

In their most recent experiments, Collinge's team infected mice by injecting their brains with the abnormal prion proteins that cause BSE in cows and vCJD in people.
The injected mice had been genetically engineered to have either the human MM genetic background or the VV background.
As expected, all MM mice developed the usual form of vCJD, as seen in humans. But only half the VV mice developed any disease at all. And the half that did develop disease had unusual damage to their brains. Instead of the usual daisy-like patterns of plaque formation seen in vCJD, the plaques were overlapping circles, like scales on a fish.
The big question is whether humans with the VV background have been developing similar plaques without anyone knowing. But the experiments suggest that if they are, the disease will either be asymptomatic, milder or too slow-developing to harm anyone.

Protected status

The other surprise came when brain material from the mice with VV disease was injected into the brains of healthy VV or MM mice. Encouragingly, VV recipients seemed immune. "None of the VV mice had clinical disease, and died of old age or other symptoms," says Wadsworth.

But as well as developing vCJD, the MM mice injected with the infected material also developed a form similar to that seen in humans with “sporadic” CJD - the most common type that is not linked with mad cow disease.
This could happen in humans only if an MM individual was accidentally infected with material from someone with the VV background, perhaps through inadequate sterilisation of surgical instruments used in brain operations. But the implication is that if this form is in humans, it would have been mistaken for classical, sporadic CJD.
The results also leave unanswered the question of how BSE and vCJD manifest themselves in MV individuals, who make up half the population.
Wadsworth says that while experiments on MV mice have been completed, the data is still being analysed. However, he says that MV individuals are the most protected against all other human forms of CJD. This so-called heterozygote genotype is most resistant even in human diseases closely related to vCJD such as Kuru.
James Ironside, head of the Medical Research Council’s CJD Surveillance Unit in Edinburgh, says that care should be taken in extrapolating the results to humans. But he believes it makes sense that BSE might cause more than one form of CJD in people. “**We already know that there are at least six different types of sporadic CJD,” he says.**
Journal reference: *Science* (DOI: 10.1126/science.1103932)

BSE researcher receives top Swiss science prize; 21 September 2004

Zurich-based scientist Adriano Aguzzi is to receive the 2004 Marcel Benoist Prize for his work on degenerative neurological diseases.
The award is presented annually for a discovery or field of study that has made an outstanding contribution to Swiss science.
The prize will be officially handed to the 44-year-old Aguzzi at a ceremony in Zurich on November 11.
Aguzzi is a neuropathologist who has conducted research into protein-related illnesses such as Creutzfeldt-Jakob Disease (CJD) – a fatal brain disorder that may be linked to the eating of cows infected with BSE, also known as mad cow’s disease.
Responding to news of the award, Aguzzi said he was grateful for the recognition but stressed that his research was “always about teamwork”.

He added that he had not yet decided how to spend the SFr100,000 ($79,000) in prize money which comes with the award.

The Marcel Benoist Foundation said Aguzzi had been selected to receive the award in recognition of his efforts to improve understanding of CJD.

Aguzzi and his team of researchers have studied the transmission of proteins to the brain in both CJD and BSE.

The Swiss interior minister, Pascal Couchepin, congratulated Aguzzi on his award during a meeting in Bern on Monday.

In a statement, the interior ministry said his research had broadened scientific knowledge about a variety of degenerative diseases, including Alzheimer’s.

Britain: new findings point to larger outbreaks of vCJD “mad cow disease”; 18 August 2004

UK scientists are upwardly revising their estimates of the number of people likely to die from new variant CJD (vCJD, also known as “mad cow disease”). It follows the death of a second patient, who contracted the disease after a blood transfusion [1].

The patient, who had received a blood transfusion from a donor diagnosed with vCJD in 1999, died five years later of a ruptured artery. An autopsy revealed that the patient was infected with vCJD in the spleen and lymph nodes.

The first case of a person being diagnosed with vCJD after a blood transfusion occurred last year, prompting an investigation that revealed another 17 people had received blood components from donors later diagnosed as carrying vCJD.

New variant CJD affects the functioning of the brain, causing personality change, loss of body function, and eventually death. It is believed to have arisen due to people eating meat infected with bovine spongiform encephalopathy (BSE). The BSE epidemic amongst cattle in the UK reached its peak incidence in January 1993 at almost 1,000 new cases per week.

BSE and vCJD are thought to be caused by a rogue form of proteins called prions. All previous cases of the disease have been in people with a particular set of markers on their prion proteins. People with this genetic make-up comprise one third of the UK population. However, prions in the latest case of the disease have a different set of markers, and more than half the population shares these.

In Britain itself, 147 mainly young people have died of vCJD. It was previously expected that this would rise to as many as 3,800. This estimate was derived from examinations of appendix and tonsil tissues removed from 12,674 people during routine surgery, of which three were found to contain abnormal prion proteins. However, this estimate assumed that only those with the less common set of markers were susceptible. The latest case has proven that a larger proportion of the population are susceptible to the disease than had previously been acknowledged.

Scientists expect that people with any of the three possible sets of markers will eventually prove susceptible to vCJD, but the incubation periods (the time during which the symptoms are not visible) are likely to be different for each set.

The Conservative government at the time of the BSE epidemic, and the Labour government that followed it, both limited their response to the danger of vCJD to ordering the removal of the brain and spinal cord from slaughtered cattle, rather than removing all UK beef from the food chain. They did this on the grounds that consuming the blood and muscle of infected animals could not transfer the disease.

There is now strong evidence that this assumption was also false. Although the amount of infective agent in the blood is much smaller than in other parts of the body, it cannot be regarded as safe.

The Blair government has been forced to acknowledge this by banning people who have received blood transfusions from donating blood themselves, so as to reduce the risk of vCJD being spread by this route. This latest measure is in addition to the removal of white blood cells from all blood used for transfusions, and the importing of blood products such as clotting agents and plasma. At present, there is no blood test to determine whether someone has the disease.

Professor Ironside, head of the CJD surveillance unit in Edinburgh that researched this latest case of vCJD said, “This finding had major implications for future estimations of numbers of vCJD cases in the UK.... A very lengthy incubation period might explain why no clinical cases of vCJD have yet been observed in this subgroup.”

He said that there could be other people in the subgroup (i.e. the group of people with the same set of markers on their prions) carrying the disease without being aware of it. They could still infect others with vCJD, via blood transfusions, organ donations or reused surgical instruments. He added, “It’s absolutely possible that there may be a new epidemic, because the cases we’ve seen so far may only be those who are unusually susceptible or have the shortest incubation periods. I’m not in the business of scaremongering, but quite clearly the idea that this problem is on the way out is unfortunately not the case at all.”
A group of researchers led by John Collinge at the Medical Research Council’s prion unit in London believe their research with mice strengthens the evidence that eating BSE-infected beef gives rise to several types of brain disease, and that these include another kind of CJD known as sporadic CJD, which had not previously been associated with BSE.
The factors affecting one’s susceptibility to these infections are complex, and involve other genetically determined characteristics besides prion markers. The research indicates that there are at least seven genes affecting the susceptibility of mice, and Collinge said that it would be “pretty surprising” if it were not also the case in humans.
Frances Hall, secretary of the Human BSE Foundation, whose son Peter died from vCJD in 1996, commented, “The hope was that only those of an unusual genetic type would develop vCJD. Unfortunately it now looks like more people could be susceptible. It’s still too early in the day to know how many people will eventually end up with this disease.”
Note:
[1] “Preclinical vCJD after blood transfusion in a *PRNP* codon 129 heterozygous patient” (Lancet 2004; 264: 527-29) by Alexander H Peden, Mark W Head, Diane L Ritchie, Jeanne E Bell, James W Ironside.

vCJD woes ahead; 14 August 2004

The prospect of a second wave of vCJD cases caused by the BSE epidemic now seems more likely.

All the 142 deaths from the human form of BSE in the UK have so far occurred in people with a set of genes that seems to make them susceptible to the disease. But last week it was **reported that someone with a different genotype had been found to be incubating vCJD after dying of other causes (The Lancet, vol 364, p 527). Half of the population has the same genotype, so the discovery is ominous.**

"**What this finding indicates is that the largest genetic subgroup is susceptible to this infection," says James Ironside,** director of the UK's National CJD Surveillance Unit in Edinburgh, UK, and a member of the study team. "From one case you can't extrapolate too far but it seems that many of them are likely to be infected."

Based on the number of cases so far, epidemiologists have estimated that there will only be another 50 to 60 cases of vCJD, though initial findings from a survey of tonsils put the figure at 3800. The latest case suggests the toll might be even higher.

When proteins attack; 07 August 2004

Humans are supposed to be protected from animal diseases by something called the species barrier. So what went wrong with BSE?

The BSE epidemic in the UK was an unmitigated disaster. It devastated the beef industry, destroyed peoples' livelihoods, unleashed a terrible disease upon the population and cost billions of pounds of public money.

But to a small group of biochemists there was a silver lining. BSE and its human equivalent, vCJD, belong to an obscure group of diseases called prion diseases, transmitted through an implausible mechanism involving shape-shifting proteins that formed clumps in the brain. Without BSE, prion science would probably have remained a backwater. Once the disease struck, however, it went mainstream.

Now, as the US ramps up its BSE testing programme and the threat of a global epidemic becomes ever more real (see "American nightmare"), prion science has come of age. At the time of the UK outbreak there were multiple unknowns and controversies - not least over the very nature of prions themselves. Today, new insights ...

Out of sight, out of mind?; 7 August 2004

Chances are you've never heard of the Garden State Racetrack in Cherry Hill, New Jersey, still less actually been there. But for the punters who frequented the track until it closed in 2001, the name looms large -- especially if they ever ate there. Because according to a local campaigner, beef served at the racetrack's restaurant is the source of a cluster of the human form of mad cow disease.

It's a bold claim, and the official response has been scathing: **no one in the US has ever been formally diagnosed with variant CJD, the human form of BSE. But back in December the US declared its first case of BSE in cattle,** and it now looks as though the disease has been circulating in the national herd for several years. That means it is possible that humans have been exposed to infected meat, which is how people are thought to catch vCJD.

Of course, these facts don't add up to the conclusion that the Garden State cluster is real. In fact, the best available evidence suggests racegoers have little cause for alarm. **But the "cluster" raises a wider question: if Americans did start catching vCJD from infected beef, would it be detected? Perhaps not. Critics say US surveillance systems for the disease are inadequate -- in many states doctors are not obliged to report suspected cases. And even when they do, those caused by infected beef could easily go unrecognised. The chilling conclusion is that the US could already be incubating a human epidemic of BSE without knowing it.**

Mad cow disease, caused by "rogue" prion proteins accumulating in the brain, emerged in the UK in 1986. It stemmed from feeding cattle the remains of other cattle, allowing each sick cow to infect numerous others. In the late 1980s the UK banned this practice and took other measures to eliminate BSE.

It was too late to stop the disease jumping to humans. In **1996 the national surveillance centre found a new form of Creutzfeldt-Jakob Disease. This disease had been known for a long time, occurring at a rate of about one case per million individuals a year. The commonest form, sporadic CJD, affects only the elderly**, and probably results from chance mutations. Post-mortems reveal characteristic clumps of protein in the brain.

But the new form was a different proposition: it affected young people and produced distinct protein clumps**. It is now generally accepted that this vCJD is a result of eating BSE-infected beef. So far there have been 142 confirmed cases in the UK.**

Since 1997 the National Prion Disease Pathology Surveillance Center at Case Western Reserve University in Cleveland, Ohio, has been on the look-out for vCJD in the US. The centre has had 1277 suspected cases of CJD referred to it, of which 792 were confirmed as

CJD. Only one was vCJD: a woman who had been brought up in the UK, and is assumed to have caught the disease there. She died in June.

But the centre can only investigate the cases it knows about. In most European countries and Canada, CJD is a notifiable disease, which means doctors are legally obliged to report all cases. In the US only 25 states have a notification policy, so cases in other states could slip through the net. The surveillance centre has sent out 60,000 letters nationwide to physicians, neurologists, pathologists and anyone else who might help, asking them to report suspected CJD cases. "We made a tremendous effort to increase awareness," says Pierluigi Gambetti, the centre's director.

The measures have helped. In 2003 there were 284 referrals, compared with just 94 in 1998. But even now only about 60 per cent of cases where CJD is listed on the death certificate as a possible cause are referred to the centre. **And some doctors may not even realise CJD is a possible cause of death.**

These concerns that vCJD could, in theory, be overlooked lend at least some plausibility to the Garden State racetrack claims. **Janet Skarbek, an accountant from New Jersey with no scientific training, took it upon herself to investigate after a close friend, aged 29, died from a degenerative brain disease four years ago. Skarbek traced 15 other individuals who had met a similar fate. The common thread was that they had all been to the racetrack and eaten at the restaurant between 1988 and 1992. And their deaths had hallmarks of CJD.**

After sustained lobbying by Skarbek, the New Jersey Department of Health and Senior Services and the Centres for Disease Control and Prevention (CDC) in Atlanta, Georgia, carried out an investigation, which included autopsies and analysis of stored brain samples from at least half the cases. In May they announced their findings. Three of the 16 people who had died, including Skarbek's friend, had not had CJD. Eleven did have CJD, but the sporadic form. This is no more than would be expected among the number of people attending the race track over that time. The other two cases, as well as one possible additional case, are still under investigation. Unfounded fears

"The cases are spread out over 12 years, and that's not a cluster," says Larry Schonberger, the CDC's coordinator of CJD surveillance. "People who've been to that racecourse are now unnecessarily fearful about their health."

But these results cannot rule out a link with BSE. **In February, Salvatore Monaco and his colleagues at the University of Verona in Italy discovered a new form of BSE in cows that produces brain disease patterns very similar to those of sporadic CJD (Proceedings of the National Academy of Sciences, vol 101, p 3065)**. And in the UK, researchers led by John Collinge at the Medical Research Council's Prion Unit in London have carried out studies in mice that are also worrying. They showed that mice injected with prions that usually cause vCJD can develop brain abnormalities typical of sporadic CJD (The EMBO Journal, vol 21, p 6348). Collinge told New Scientist that his more recent research, still unpublished, suggests it is the strain of the mouse that determines which disease appears. "The implication is that some people exposed to BSE might get disease with pathology which we recognise as sporadic CJD," he says. He warns, however, against jumping to conclusions. "You must be cautious extrapolating from animal models."

The number of sporadic CJD cases in the UK has risen since surveillance began, from 28 in 1990, to 50 in 2000. Some of that is no doubt due to better reporting systems. "The question is whether there's a minority of cases which have another cause," Collinge says. His team is now trying to develop molecular probes capable of identifying a subtype of "sporadic" CJD that may in fact be human BSE.

Perhaps unsurprisingly, Skarbek has jumped on these findings. She has asked Collinge to analyse some of the brain samples, and is lobbying the CDC to carry out a full epidemiological investigation that would delve into the life histories and eating habits of the racetrack goers. The CDC, however, has no plans to comply and denies any suggestion of a cover-up. "There's no conspiracy," Gambetti says. "On the contrary, we're always on the watch for something unusual. If there are cases out there, we want to detect them."

American nightmare; 7 August 2004

The US has so far found only one mad cow, but experts say there's no such thing as an isolated case. So where are the rest?

At VERN'S Slaughterhouse in Moses Lake, Washington, Tuesday 9 December 2003 was a busy day. "We were swamped," recalls slaughterman Dave Louthan - especially with "downers", sickly cows too weak to stand.

Amid the rush, one cow caught Louthan's attention - a white, 6-year-old Holstein standing in a truck at the downers' entrance. She was causing trouble, and Louthan admits he should have taken her round to the pens where cattle that could walk awaited slaughter. "But I was in a hurry," he says, "and she didn't want to step down off the trailer." Before she could trample the other cattle, "I put a hole in her forehead".

At that time the US Department of Agriculture was paying slaughterhouses for brain samples from downers, to test for BSE. The Holstein was officially among the downers, so Louthan took a sample. Two weeks later, the USDA announced that the cow was ...

Fears of vCJD timebomb revieved; 6 August 2004

A “significant number” of people in the UK may be harbouring vCJD with no clinical symptoms, reveals the study of a new case. The work suggests the epidemic of the human form of mad cow disease may be far from over.

The second case of vCJD linked to a blood transfusion was announced by the UK department of health on 22 July. **But investigations have now revealed that this person, who had no clinical symptoms and died from other causes,** is the first case of vCJD to be found in the largest genetic subgroup of the population.

The discovery has major implications for the ultimate extent of the vCJD epidemic, as about 50 per cent of the UK population falls into this group.

“What this finding now indicates is that the largest genetic subgroup is susceptible to this infection,” warns James Ironside, director of the UK’s National CJD Surveillance Unit in Edinburgh, UK, and one of the study team. “From one case you can’t extrapolate too far but it seems that many of them are likely to be infected.”

This is because exposure to vCJD by eating infected beef was “likely to be widespread” in the late 1980s and early 1990s, he says. The 142 deaths seen in the UK so far have affected people in a smaller subgroup, who may have a shorter incubation time.

Person-to-person

The case is also ominous because it is the second believed to be acquired through a person-to-person blood transfusion. Furthermore, the donor had not developed any symptoms of vCJD at the time of donation, showing such people can still transmit the disease.

The first transfusion case was identified in December 2003 in the UK. The patient died from vCJD seven years after receiving a blood transfusion from a donor who also later died of the disease.

"I don't want to be alarmist about this – but this really is an important case," Ironside told "Firstly, it means this genotype is susceptible; secondly, it clearly means vCJD is being harboured; and thirdly, the illness was almost certainly acquired as a result of a blood transfusion."

He says the "secondary transmission" via blood transfusion also raises the issue of transmission via surgical instruments. No known cases of vCJD have been transmitted this way, but Ironside notes: "Unfortunately, with this disease, if the worst can happen it apparently does."

Common combination

In the latest case, the patient died from a non-neurological disorder five years after receiving blood from a donor who subsequently developed and died from vCJD. The patient was the one of 17 identified as having received blood from infected donors.

An autopsy revealed that the patient had the mutated prion proteins responsible for vCJD in the spleen, despite showing no symptoms of the devastating disease. These rogue proteins were not found in the brain, spinal cord, or tonsils.

Crucially, the patient is the first to have the commonest combination of genes for the normal prion protein. All vCJD deaths to date have occurred in people who have two identical genes – one each from their mother and father. But there is another version of the gene and the latest patient was "heterozygous", i.e. they had one of each type.

Ironside says that because the number of vCJD cases peaked in 2000, "there was a view that may be this was the beginning of the end". But the new case shows that heterozygous people may just be incubating the disease for longer.

That view is supported by studies of other similar diseases, like kuru in Papua New Guinea, which show that heterozygous people can incubate the diseases for a long time. "Incubation periods over several decades are not uncommon," says Ironside.

Tonsil and appendix

Azra Ghani, an epidemiologist at Imperial College London, UK, who has been modelling the course of the vCJD epidemic agrees the discovery is of major importance. She estimates that if heterozygous people are similarly susceptible to those who are not, there might be a doubling of projected deaths.

Her group currently estimates 50 to 60 more cases in the UK by 2080, based on the numbers of real cases. **However, she cautions that initial data from a survey of tonsil and appendix tissue suggests there may be 3800 more cases.**

She also told that the potential for blood transfusion to spread vCJD holds "greater uncertainty" as there are many unanswered questions: **"How infectious is blood? What sort of dose would you need?"**

Unquantifiable risk

The UK's Department of Health stepped up its precautionary measures for blood transfusion in December 2003 and again after the second case was announced in July.

John Reid, the secretary of state for health, stressed the “small but unquantifiable risk” at that time. "People should continue to have a blood transfusion when it is really necessary. Any slight risk associated with receiving blood must be balanced against the significant risk of not receiving it,” he said.

Ironside stresses the urgent need for a blood test to detect vCJD. If many people are incubating the disease, he warns, “we have no means of detecting them preclinically”.

“We should take some comfort from that fact that we have not seen a huge mass of epidemics in humans,” he says. “But the idea that vCJD’s going away now cannot be considered to be the case.”

Journal reference: *The Lancet* (vol 364, p 527)

BSE/Mad Cow Disease crisis provokes trade war; 2 August 2004

The spread of Bovine Spongiform Encephalopathy (BSE), or mad cow disease, has provoked a trade war in cattle and beef products.

The World Organisation for Animal Health (OIE, Organisation Internationale des Epizooties) has blamed the “recent international trade disruptions” on “the apparent misinterpretation of BSE standards or the failure to implement these standards” by governments involved in the cattle and beef trade.

The OIE has issued standards designed to reduce the spread of BSE via trade using a risk assessment that counts the number of BSE cases found in a country and the controls used to prevent the spread of the disease. There are five categories of risk ranging from “free of BSE” to high risk. Governments have sought to block imports of cattle and beef by declaring importing countries high risk.

BSE in cattle has been linked with the fatal human brain condition variant Creutzfeld-Jacob Disease (vCJD), which has affected at least 147 people, the vast majority of whom lived in the United Kingdom where the disease originated. Recent research on tonsils and appendixes suggests that as many as 3,800 people in the UK may be harbouring vCJD but there are higher and lower estimates.

The total number of confirmed cases of BSE in cattle in the UK since its discovery in 1986 is around 180,000. Currently the UK is in the highest risk category, but the British government asked the European Commission in June 2003 to downgrade its status to moderate risk. Ben Bradshaw, Minister for the Department for Environment, Food and Rural Affairs, claimed the reduced status was justified because there were only 374 BSE cases in 2003 and tough control measures are in place. He said, “If we are successful and enter the reduced category this will be a major turning point as it will bring us in line with a number of other member states making trade much more accessible.” It will “enable us to compete again with much of Europe by re-opening important export markets for British beef,” Bradshaw added.

The beef industry could be worth £11 billion in exports.

The European Food Safety Authority agreed in May 2004 to the moderate risk status, but its implementation is being delayed by public health agencies.

Controls in the UK involve a ban on recycling mammalian-based feed to farm animals; a ban on cattle over 30-months-old and all high risk animal tissues (Specified Risk Materials SRM), like the brain and spine, from entering the human food chain; and the introduction of a cattle tagging and tracking system.

Besides seeking “moderate risk” status from Europe, the British government is also trying to relax the ban on the consumption of beef from cattle over 30-months-old. Robert Forster, chief executive of the National Beef Association, has complained that the scheme costs about £360 million a year “to incinerate perfectly good beef”. The government-run British Food Standards Agency says that the Department of Health should lift the ban and replace it with a

BSE test on every cow, even though the government's own expert Spongiform Encephalopathy Advisory Committee warned the decision was based "on expert judgment rather than being fully informed by all the required data".

This pressure for change comes about despite concerns that the controls are flouted and that there is worrying new scientific evidence about the nature of BSE.

A recent report by the Public Accounts Committee of the UK Parliament found that the Cattle Tracing System was "developed in haste and has suffered from serious technical difficulties" which has resulted in £14 million fines from the European Commission and a possible further £36 million to pay. Two thirds of the 700 staff employed by the CTS service are currently correcting 1.2 million "anomalies" involving 200,000 cattle and two million sheep movements. Committee chairman, MP Edward Leigh, said the CTS "does not fully meet the needs of state veterinarians to control outbreaks of infectious diseases amongst cattle, which is all the more unacceptable given that it was introduced in response to the BSE crisis in the 1990s."

Of more concern are reports from scientists that there may be different types of BSE-like diseases in cattle that could produce other fatal brain diseases. Italian researchers have identified a brain pattern in two cows that is different from the traditional BSE one and French and Japanese scientists have also discovered atypical cases. In the UK, Professor John Collinge has injected mice with BSE-infected material and whilst some developed a vCJD response, others resembled sporadic CJD, a related disease that affects older people. When asked why British scientists had not found different types of BSE previously, Collinge said it was because the government had sponsored such research. "It has always been on the cards, but it has not been a terribly popular thing to suggest," he added.

An additional worry is that the fatal sheep disease scrapie could be masking BSE. If this proves correct most of the UK flock would have to be slaughtered. The British government introduced compulsory measures in June 2004 to eradicate scrapie from affected flocks. At present dead sheep are tested for scrapie and those found positive are also tested for BSE. However, there have been 78 unconfirmed test results that do not resemble either BSE or scrapie.

Although the BSE crisis has affected British farming most severely, it has also hit trade worldwide, including Japan and North America.

In Japan, the discovery of BSE in September 2001 led Health, Labour and Welfare Minister Chikara Sakaguchi to introduce BSE testing on all slaughtered animals. The consumption of beef, including imported meat, fell as a result. In December 2003, Japan banned American beef after the first confirmed case of BSE in the US was found in a cow in the state of Washington. Although the US domestic market for beef has not declined, exports have been hard hit. Trade with Argentina, Uruguay and Paraguay for example has dropped from $7.5 billion to $4.8 billion.

The Japanese government has demanded that the US authorities test all slaughtered cattle for the disease or provide similar safety assurances before US beef imports can start again. The US government has rejected this request, claiming that blanket testing of all cattle is unscientific.

Testing in the US is only carried out on cattle showing signs of central nervous stress, although cattle can incubate the disease for many years before it shows itself. In 2003, the US Food and Drug Administration (USDA) carried out tests on only 20,000 cows out of the nearly 40 million head of cattle slaughtered.

The US government's main response to the BSE crisis has been to rely on the removal of SRMs from animal feedstuffs—introduced in 1997. Following the discovery of BSE in Washington state the USDA banned SRMs from meat intended for human consumption in

January 2004 and is considering their prohibition in cosmetics and dietary supplements. An initial $19 million voluntary National Animal Identification System is also being considered.
Sakaguchi has so far refused to bow to Washington's demands to withdraw his ban. He said, "We have to think about people's feeling as well as scientific issues. We will examine whether we can provide data that reassure consumers." He added, "I made a decision on (introducing) the blanket testing. I am opposed to a hasty review."
Whilst the US government has criticised the Japanese government for banning US beef imports based on a single BSE case (and has no doubt put pressure on the OIE to issue its statement), it has restricted imports from its neighbour Canada following the discovery of BSE there in May 2003.
The US-Canadian border is closed to live cattle and beef cuts from over 30-month-old cows. Prices have crashed in Canada and thousands of workers have lost their jobs. Exports have slumped from $4 billion to $1.5 billion.
In Alberta, home to one-third of Canada's cattle and where two-thirds of the country's beef is produced, over half the municipal districts have declared themselves economic disaster areas because of BSE. Alberta Agriculture Minister Shirley McClellan said she did not know when the border would reopen and called on the US government to resist closing it again if more cases of BSE are found. "We must ensure countries cannot use an incident like this as a trade barrier, that decisions are really based on sound science," said McClellan.
Recently, Canadian Prime Minister Paul Martin blamed US "special interests" for blocking free trade between the two countries and explained that Canada is considering building large processing plants to cope with the oversupply of cattle, targeting large US markets such as South Korea.
The Canadian government has also been quick to silence scientists who have a "special interest" in maintaining public health. Three senior scientists who worked in the veterinary drugs approval laboratories of Health Canada—Shiv Chopra, Margaret Haydon and Gerard Lambert—were sacked on July 14 this year. The three had repeatedly criticised Health Canada policies in public, claiming they were pressured into approving drugs they thought might endanger humans. Long before BSE was detected in Alberta they had warned their managers that the department's policies to fight BSE were inadequate.
The BSE and vCJD diseases have become an international problem. However, the division of the world into competing nation states and the subordination of social needs to the profits of huge multinational corporations had obstructed an internationally coordinated effort to overcome it. Instead, it results in economic trade war. As long as the production of humanity's food is governed by the drive to maximise profit, preventable diseases like vCJD quickly escalate into public health disasters.

Synthesis marks prion disease breakthrough; 29 July 2004

In a breakthrough that could offer new ways to combat mad cow disease and related human brain disorders like vCJD, the infectious prions which trigger these diseases may have been synthesised in a laboratory for the first time.

Researchers have been trying - unsuccessfully - to create mammalian prions in a test tube every since Stanley Prusiner, a biochemist at the University of California, San Francisco, first theorised in 1982 that prions were infectious particles composed entirely of protein.

Now Prusiner's team reports tantalising evidence that they have been able to build a mammalian prion from scratch. When these synthetic prions were injected into the brains of mice, they triggered a prion disease that could be passed to other animals.

"The implications are huge and open up many new ways to study prions and gain new insights about prion disease," says Giuseppe Legname, a senior scientist in Prusiner's lab and one of the lead authors of the study.

"This is an exciting and fascinating initial experiment," says Byron Caughey, a prion researcher at the Rocky Mountain Laboratories in Hamilton, Montana. "But we're on new ground here. There are details that need to be worked out before it's clear this is definitive proof."

Corrupted shape

According to Prusiner's protein-only prion theory, BSE and related diseases occur when a protein called PrP found in healthy brains assumes a corrupted "prion" shape. The prion can then perform an odd sort of replication by coaxing healthy copies of PrP to flip into the diseased shape.

Folding purified PrP into the prion form in a test tube has been the holy grail of prion science because it seemed like the only way to silence all doubt that protein - and not some contaminating brain virus, for example - were actually responsible for BSE. Furthermore, being able to study prions in isolation would give new insights into how PrP folds into a prion and how to prevent or reverse the process to treat prion diseases.

The protein-only theory received a boost in 2000 with the discovery of other proteins from yeast that seem to form prions in a test tube (August 2000). But despite exhaustive efforts by many labs, no one had managed to perform the same trick with ordinary PrP.

Instead, Legname says his team used a smaller fragment of PrP that was believed to form prions more easily. And rather than inject the synthetic prion into ordinary mice, the researchers used animals that were genetically modified to produce the same PrP fragment at a level 16 times higher than normal, making them more susceptible to prion infection.

The PrP fragment was produced in bacteria, purified and then encouraged to form prion containing fibres known as amyloid.

Rigid tails

Control mice that received a brain injection without the lab-made prions did not develop prion disease after 670 days. But animals that received the synthetic prions started showing the wobbly gait, ungroomed fur and rigid tails that are the clinical signs of rodent prion disease after 380 days.

Extracts from the brains of those diseased animals were injected into normal mice which started getting sick after, on average, only 154 days. That suggested the starting number of synthetic prions was low, but improved after one cycle of replication in a mouse brain, says Legname.

The data suggests that PrP did form a prion in the test tube. But Caughey points out an alternative, though he admits, less likely explanation: the prions weren't created in the test tubes, but in brains of mice overproducing the protein.

If that is true, those prions must have replicated so slowly they do not normally kill the mouse, but the additional injection of PrP accelerated their replication. To eliminate this possibility, it will be necessary to show that brain extracts from uninjected mice never trigger prion disease even when injected into a new animal.

Legname says those experiments are already under way. Prusiner's team is also busy testing many PrP folding conditions to find ways to make prion formation in the laboratory much more efficient.

Journal reference: *Science* (vol 305, p 673)

BSE downer; 17 July 2004

In a surprise move, the US has postponed long-awaited plans to ban material from animal feed that might be infected with BSE. It is the second time the US has backed away from tougher feed restrictions this year, and new rules are unlikely before the presidential election in November.

In January the US Food and Drug Administration announced that it would ban cattle blood and other potentially infected material from cattle feed, but postponed the move after an international scientific panel recommended more stringent measures. **Now the FDA is considering banning all meat meal except fish from cattle feed and banning cattle brain and other high-risk "SRM" tissues, and sick or "downer" cattle, from chicken and pig feed, because these could contaminate cattle feed.**

But instead of implementing these measures as expected, the FDA announced on 9 July that it would wait, asking for "comments and scientific information" on the proposals, all of which are watered-down versions of the measures Europe needed to control BSE. "The FDA does not need another round of comments," Jean Halloran of the US Consumer's Union claimed. "They know what needs to be done." **But the American Meat Institute, an industry group, greeted the news by repeating its opposition to banning SRM from feed.**

The shapeshifters;17 July 2004

Prions cause havoc when things go wrong. But a controversial theory suggests that behind the scenes, prion-like proteins are keeping our bodies ticking over

Pity the poor prion protein. If ever biology had a case of "give a dog a bad name", this is it. **Prions are known almost exclusively for their role in BSE, CJD and other prion diseases. Their principal claim to notoriety is the ability to morph from an inoffensive shape into a rogue one that causes other prion molecules to follow suit, setting off a deadly chain reaction.**

But perhaps it won't be long before this most maligned of molecules enjoys a change of image. A number of scientists are starting to believe that the ability to shape-shift might not be so sinister after all. For example, a Nobel prizewinning neuroscientist has stumbled upon a prion-like protein that, instead of causing disease, might change shape in order to play a vital function in memory (see "Memory makes its mark"). And a biochemist thinks he has uncovered a whole family ...

Second US cow tests positive for BSE; 28 June 2004

A second cow in the US has produced an initial positive result for BSE. US officials are saying it is “very likely” the cow will turn out to be negative. However, in most countries, the test used is wrong about a positive result only about once in a thousand times.

The US Department of Agriculture announced the result late on Friday. It was the first positive test since the US began testing thousands of cattle for BSE on 1 June. The testing is in response to the discovery of its first mad cow in December 2003.

However, the USDA is calling the result "inconclusive" until it is confirmed by immunohistochemistry (IHC) at the US's National Veterinary Services Laboratories in Ames, Iowa.

IHC is a highly reliable but slower BSE test. In a briefing on Friday, John Clifford, deputy head of the US Department of Agriculture's Veterinary Services Programme, said the IHC results will take four to seven days.

The USDA will only give the location of the animal was and why it was tested if that result comes back positive, said Clifford. The USDA is targeting animals that show BSE-like symptoms, or which die for unknown reasons, or are unable to stand. Such "high risk" cattle are 10 to 15 times more likely to have BSE.

Extremely sensitive

But Clifford stated repeatedly that "it's very likely this animal could be negative". He stressed that the BioRad test used was "designed to be extremely sensitive" to catch any possibly infected animal, some of which "will end up negative during further testing".
Asked the odds of the result being a false positive, Clifford said "we wouldn't want to provide that type of information". But the rate of false positives for the BioRad procedure was measured by the European Commission when it evaluated BSE tests for use in the EU in 2003.
Those data have never been published, but industry sources who have seen them say the BioRad test had a false positive rate of about one in a thousand initial tests, a rate borne out subsequently in practical experience. BioRad is used in Germany and Belgium.
In Europe and Japan, if a cow's brain initially tests positive with BioRad, it is tested twice more. Only if one of those repeated tests is also positive is the sample sent for IHC confirmation. The false positive rate after such repeated testing is even lower, around one in 100,000 for BioRad. It is not known whether the USDA is doing this.

True incidence

However, Markus Moser of the Swiss firm Prionics, which makes a rival ELISA test for BSE, points out that if the true incidence of BSE in cattle is much lower that the test's false positive rate, most of the positive tests obtained with the test will indeed be false.
In Japan, where the BSE incidence is just one tenth of the BioRad false positive rate, only eight of the first 113 positive BioRad tests were confirmed.
It may be that this is what USDA is counting on when it says its positive test is "very likely" to turn out false. But no one knows if the true incidence of BSE in the US is so low – that is what the testing programme is supposed to measure

No BSE-free cow; 05 June 2004

Reports that the first cow genetically engineered to be immune to BSE will soon be born have turned out to be misleading

Reports that the first cow genetically engineered to be immune to BSE will soon be born have turned out to be misleading.

In theory, creating BSE-free animals is simple: delete both copies of the gene for the PrP protein that causes prion diseases when it becomes mishapen, and animals cannot develop the

disease. But in practice, engineering such animals is time-consuming and very costly, and past attempts to create cows that lack the gene have failed (January 2002).

So when Kirin Brewery of Japan this week announced that a cow was pregnant with a calf that lacks the PrP protein, the story received global press coverage. But the actual work is being carried out by Kirin's partner, Hematech of Connecticut, and James Robl, the company's chief scientific officer, told that so far the US company has only created cell lines lacking the ...

Cows immune to BSE near reality;1 June 2004

A major advance towards producing prion-free cows that would be immune to mad cow disease has been made by researchers at companies in the US and Japan.

Their principal aim is to make genetically modified cattle that produce pharmaceuticals in their milk. But the companies hope that also making the animals resistant to BSE (bovine spongiform encephalopathy) will reassure consumers.

The researchers have now achieved the considerable feat of creating cell lines which have both copies of the cow's PrP gene switched off. The PrP protein can be switched to an infectious state by contact with a mutated prion. This switch causes prion diseases such as BSE in cows and variant Creutzfeldt Jakob Disease (vCJD) in humans.

Making live animals from these cell lines should be relatively straightforward using cloning techniques similar to those that created Dolly the sheep.

The companies say they have no intention of producing prion-free animals destined for human consumption. Instead they want to assuage public fears about pharmaceuticals derived from cow's milk, even though the process used to extract proteins from milk has already been shown to remove prion contamination.

Consumer choice

"Because of public perception, we feel an added benefit would be to knock out the gene and remove the possibility that the animals could be infected," says James Robl, chief scientific officer at US biotech company, which carried out the research jointly with the Kirin Brewery in Tokyo, Japan. "Japanese consumers have a high degree of concern about BSE."
Some experts have suggested creating prion-free cattle to eliminate BSE from the beef herd. But it is not clear that consumers would prefer genetically modified beef over meat that has a very small chance of infection with prions. To date there have only been around 140 cases of vCJD in total worldwide, although the ultimate extent of the human epidemic remains uncertain.
Furthermore, replacing beef herds with prion-free cattle would take decades. "Getting a herd of any size would be quite difficult," says Harry Griffin at the Roslin Institute in Scotland, where Dolly was cloned.
Another question mark hangs over what effect knocking out both copies of PrP will have on cows. Mice lacking both copies appear to be normal, although some disputed research suggests they may have disrupted sleep patterns.

Low yield

Hematech and Kirin's aim is to produce cows that produce human antibodies that can be harvested from the animals' milk. These could be used to treat people with specific infections for which there are no vaccines.
In 2002, they engineered a mini chromosome carrying the human antibody gene into cows. But, because the cattle still had their own version of the gene, the yield of human antibodies was low.
They have now managed to create around 150 cow embryos which have both copies of the bovine antibody gene knocked out. These will be born in 2005, and some have also been engineered to include the human version. The companies hope they will produce a better yield of human antibodies.
The prion-free cell lines also lack both copies of the bovine antibody gene, but Hematech has yet to clone these to produce embryos.
Although sheep with one copy of the PrP gene switched off have already been created at the Roslin Institute, it has proved difficult until now to knock both genes out in livestock

Tonsil tests suggest thousands harbour vCJD; 21 May 2004

Almost 4000 Britons aged between 10 and 30 may be harbouring the prion proteins that cause the human form of mad cow disease. The new estimate comes from direct analyses of human biopsies, and is much higher than epidemiological projections of the likely number of deaths from variant Creutzfeldt-Jakob Disease (vCJD).

The investigators discovered three infected tonsil or appendix samples from a total of 12,674 stored between 1995 and 1999. However, because so few positive samples were found, the projected total of 3808 can only be speculative. Furthermore, harbouring the prions may not necessarily lead to vCJD.

"I don't think too much should be read into our findings, but they should be investigated further," says David Hilton, of the Derriford Hospital in Plymouth, UK, who led the study.

He notes that only one of the three positive samples matched the usual pattern of prion accumulation seen in confirmed vCJD cases.

The other two are different. "It could mean these are false positives," says Hilton. If they are, then the predicted incidence of the disease drops by two-thirds, from 237 per million British citizens to just 79 per million.

Fresh samples

Concern at Hilton's preliminary findings in 2002 prompted the UK government's Health Protection Agency to launch in 2003 a much larger screening study based on an archive of 100,000 freshly collected tonsil samples.
"The most important thing will be to have a very large number of samples in good condition," says Pat Troop, chief executive of the HPA. "We'll test them in batches, and if there are significant findings en route, I would expect the Department of Health to publish them."
But many of the fresh samples will come from children whose food is now free of the cow prions thought to have passed BSE to humans. "That is a possible weakness," says Hilton.
His own study focused on preserved samples from the people most heavily exposed during childhood and early adulthood to meat from cattle with BSE.

Species barrier

Hilton says that the low number of three positives from such a heavily exposed group may indicate that cow prions seldom pass from cows to humans, because of a so-called "species barrier".
"From the observed cases, the species barrier does seem to be very high," he says. "But we need to do these large scale studies to find out more."
The Department of Health says that the uncertainties in Hilton's studies justify its measures to protect patients. These include filtering potentially infective white blood cells from blood donations, and upgrading sterilisation equipment to stop prions spreading on surgical instruments.
Meanwhile, the number of recorded vCJD cases continues to fall from a peak of 28 deaths in 2000. In 2003 there were 18 deaths, with two so far in 2004. In April 2003, when the total deaths had reached 121, epidemiologists downgraded their "best guess" projection of all further deaths to just 40.
Journal reference: *Journal of Pathology* (DOI: 10.1002/path.1580)

Maverick researcher impugns prion theory; 29 *April 2004*

He thinks bacteria affect proteins, causing ills such as chronic wasting

San Francisco - More than 20 years ago, **maverick researcher Stanley Prusiner** endured ridicule when he proposed that a rogue protein molecule devoid of any DNA could replicate itself and cause a family of fatal brain disorders in people and animals, including mad cow disease.His idea was revolutionary, and in 1997 Prusiner won the Nobel Prize. **His prion theory is considered dogma by many in the field of neuropathology.**

Now, another loner is facing criticism for his own sacrilegious scientific concept: that Prusiner's prions are not the infectious agents and that the real culprit in the diseases is an unusual bacterium. In research presented Thursday, **Frank Bastian, a professor of neuropathology at Tulane University, said he has found bacteria known as spiroplasma in brain tissue samples from people and animals that died** of so-called transmissible spongiform encephalopathies, including chronic wasting disease in deer, scrapie in sheep and Creutzfeldt-Jakob disease in people.

Spiroplasma **were not found in matched brain samples of the animals and people who did not have those diseases**. Bastian said he believes that TSE diseases begin when spiroplasma attach to normal prion protein in brain cells, leading to a variety of invariably fatal brain disorders. His latest work was presented Thursday at the American Academy of Neurology's annual meeting in San Francisco. If he is right - and at the moment few scientists think he is - the prion theory may face a major setback.

At the same time, Bastian said, it could open up possibilities for early detection of those diseases and new treatments. Herds of wild deer could be given food laced with a vaccine that could halt the spread of chronic wasting disease, or a diagnostic test could identify people with Creutzfeldt-Jakob disease early enough so they could be

given a drug to stop the advance of the disorder. Such scenarios border on science fiction at the moment.

And Bastian and the few other contrarians in the field face a daunting task in proving that microbes cause the diseases. **The overwhelming majority of researchers and neuropathologists who work with TSE diseases largely have accepted the prion hypothesis: that an abnormally shaped protein, with no DNA or other genetic material, can cause normal protein in the brain to adopt its misfolded shape, leading to the irreversible death of brain cells.**

But Bastian, 65, a single-minded researcher who tends to give long, rambling answers to questions, methodically has plodded along with his controversial theory ever since he first found the presence of spiroplasma while doing an autopsy on the brain of a Creutzfeldt-Jakob patient in 1976. "Frank Bastian has been chasing spiroplasma for 20 years now," said Michael Hart, professor and chairman of pathology at the University of Wisconsin-Madison. "He's been ridiculed by the prion community. I don't think he deserves that."

Hart noted that there is intense competition for research money as well as the fame that comes with future breakthroughs."He's just threatening to other people's research," Hart said. "There's an awful lot of jealousy in the field." Already two Nobel Prizes have been handed out for TSE-related research**. If Bastian or someone else proves that a pathogen is involved, a third Nobel should be awarded, Hart said.**

Hart, the editor of the leading neuropathology journal in the U.S., himself has taken flak for publishing Bastian's work. In 2001, the Journal of Neuropathology and Experimental Neurology published one of Bastian's first studies showing the presence of spiroplasma in 13 of 13 Creutzfeldt-Jakob cases and five of nine sheep scrapie cases, but in zero of 50 control brains.

"I was really upset with Michael Hart for even publishing that," said Stephen DeArmond, a Prusiner colleague and a professor of neuropathology at the University of California, San Francisco. An overwhelming amount of research has established the prion theory, DeArmond said. There is no organism such as spiroplasma that is causing TSE diseases, he added. "Nobody else has duplicated it (Bastian's work), and nobody else in the world believes it," he said. "Right now, he is a loner." Bastian has a lot of work to do, DeArmond added. In addition to verifying the presence of spiroplasma, it has to be shown that different strains of the pathogen cause different forms of TSE disease, DeArmond said.

Bastian also has to show that purified spiroplasma, just like prions, are resistant to radiation and heat. "Stan did all that," DeArmond said. Prusiner was not available for comment. But DeArmond said he has discussed Bastian's work with Prusiner and whether his lab, one of the premier TSE facilities in the country, should research spiroplasma. Prusiner's reaction, DeArmond said, was "Why should I do that? We have our own hypothesis."

Long way from proof

Indeed, so far Bastian has shown only an association between spiroplasma and TSE diseases. He has not proved that the organism actually causes the disease.

On the other hand, evidence of the infectivity of prions also has been indirect. No one yet has been able to purify and isolate prions from infected brain tissue and show that they cause disease.

Bastian said he believes that spiroplasma attaches to normal prion protein in brain cells and causes it to misfold."I have to prove this," he concedes. But he noted that last year Japanese researchers showed that brucella bacteria, which cause brucellosis, could bind to normal prion protein. If brucella can do it, why not spiroplasma, Bastian said. "There is something else binding to that protein," he said. "Whether I'm right or wrong (about spiroplasma), we should be going in a different direction."

Spiroplasma are very small pathogens, roughly about the size of some disease-causing viruses. They have no cell wall and are resistant to heat and chemicals. Spiroplasma are found in many insects and plants. The coiled microbes "move around like jack-in-the-boxes," Bastian said.

Prion theory skepticism

Bastian is one of a small group of scientists who remain skeptical of the prion hypothesis**. Laura Manuelidis, professor and head of neuropathology at Yale University Medical School, said the most likely explanation for TSE diseases is a stealthy, slow-acting virus.** "I don't believe protein is infectious," she said. "I think it's as likely to find infection with that protein as it is to find weapons of mass destruction in Iraq."

Manuelidis said, so far, Bastian has not provided convincing proof of his theory. But his point of view deserves to be explored, she said. "I just find it odd that everybody who writes something different is ridiculed," she said. "That's what they do in this field. It's stingy, small and graceless."

In recent years, a small but growing number of scientists have raised questions about the prion theory, said Bruce Chesebro, a leading TSE researcher with the National Institutes of Health. Chesebro said he believes that eventually the real infectious agent will be found to have DNA. **Spiroplasma is a "tantalizing idea," but Bastian has not published enough research to be convincing, Chesebro said.** "I don't think it's close to being proven," Chesebro said.

As scientists have done for decades, Bastian on Thursday stood by a blown-up copy of his findings that was posted to a bulletin board in a room filled with dozens of other scientists standing by their own posters. A Japanese researcher walked by and quizzed him about his work. They exchanged e-mail addresses. The Japanese researcher then took a digital picture of Bastian standing next to his poster.

"In the past 25 years, I haven't seen anything that has dissuaded me from this," Bastian said. "I'll work on this until I die or until we solve it. "I'm having fun."

Mysterious BSE-like disease found in sheep; 8 April 2004

A massive research programme to find out whether BSE is circulating in British sheep has turned up its first suspicious result. But while scientists say the sheep did not have conventional BSE, they cannot rule out the possibility that it could have had a new form of mad cow disease that has adapted to sheep.

Britain's Department for Environment, Food and Rural Affairs has announced that the Veterinary Laboratories Agency in Weybridge, England, had found "a type of scrapie not previously seen in the UK". Scrapie is a sheep disease similar to BSE which is not generally thought to harm people.

DEFRA said the disease-causing prion detected in the sheep's brain "had some characteristics similar to experimental BSE in sheep", but that on other tests it resembled neither BSE nor "previously recognised types of scrapie".

The UK's Food Standards Agency said in a statement: "Uncertainties still remain on this issue. However, based on the best scientific evidence to date, we are not advising against eating lamb and sheep meat."

Meat and bone

There have long been fears that sheep which ate cattle-derived meat and bone meal during Britain's BSE epidemic in the 1980s might have acquired BSE, although they have never been confirmed.

Unlike BSE in cattle, prion diseases spread directly from sheep to sheep. So any BSE in sheep could still be circulating despite subsequent bans on animal-derived feed.

Furthermore, sheep experimentally fed BSE develop a disease indistinguishable from ordinary scrapie, making detection very difficult. Yet the prion from such animals still behaves like BSE, and could cause the fatal human disease vCJD.

Worse, sheep carry prions in more tissues than cattle, including the muscle that people eat, so BSE-infected sheep could cause more human disease than mad cows.

A previous attempt to determine whether British sheep had acquired BSE went spectacularly wrong in 2001 when sheep and cattle brains were mixed up in the lab. But since then, the VLA has tested the brains of all 1019 newly reported cases of scrapie, as well as 1125 scrapie brains dating back to 1998, with tests designed to distinguish scrapie from BSE.

Blot test

The new result announced on Wednesday, from a sheep recently reported with scrapie symptoms, is the first to give results that resembled BSE. Danny Matthews of the VLA told New Scientist that in a prion test called a western blot, the sheep's brain did not bind an antibody called P4. P4 also does not bind prions from sheep experimentally infected with BSE, but does bind all but one forms of scrapie tested with it.

Also like BSE, the form of the prion without a sugar attached to it had a lower molecular weight than the form found in scrapie. But the ratio of prions with different numbers of sugars on them looked like scrapie, not BSE, says Matthews.

Most conclusively, immunohistochemistry (IHC), in which thin slices of the sheep's brain were stained with various antibodies, showed prions had accumulated in different parts of the brain and different kinds of cells from BSE - or any known form of scrapie.

Passing change

IHC seems to be a reliable indicator of BSE, as it has given a constant pattern in the 100 sheep of different genetic varieties experimentally infected with BSE and tested so far. But so little scrapie has been tested, says Matthews, it is not known if one strain might give these results on the tests.
The IHC pattern reliably indicates BSE, says Matthews, having been constant in the 100 experimentally infected sheep of different genetic varieties tested so far. But so little scrapie has been tested, he says, it is not known if one strain might give these results on the tests.
One possibility, he says, is that the sheep might have been carrying a prion initially derived from BSE. Passage into new species is well known to change prions.
BSE from experimentally infected sheep has so far been passed to just one more round of sheep, with no apparent change. "But we don't know if passage through many sheep, of different genetic types, might change it so it no longer gives the same pattern in IHC or western blots," says Matthews. "Those experiments are underway now."
Any such new incarnation of BSE in sheep may - or may not - have lost its ability to harm humans.

US to test many more cattle for BSE; 16 March 2004

The US is to follow the advice of foreign scientists and test up to half a million cattle for BSE, in order to determine the prevalence of the deadly disease in the national herd.

The new policy means five to 10 times more cattle will be tested than previously planned. The change is in response to an international panel of scientists who strongly advised the US in February that it had to test far more cattle if it wanted to measure the extent of BSE infection.

However, the US has not yet acted on the scientists' other recommendations, notably to stop feeding all chicken and ruminant remains to cattle, and to keep high risk cattle tissue, such as brain, out of animal feed as well as human food.

The first known case of BSE in the US was found in Washington state in December. Since then, the US Department of Agriculture had stuck to its previous plan to test 40,000 cattle in 2004.

But on Monday, Ron DeHaven, the US's chief veterinary officer, said that in the 12 to 18 months from July, the USDA would instead test "as many as possible" of the estimated 446,000 cattle each year that are lame, unable to stand, found dead, or have neurological symptoms.

Downer animals

This is precisely what was recommended by the international panel of scientists, which was asked for its advice by the USDA and included BSE experts from New Zealand, the UK and Switzerland.
DeHaven said testing between 201,000 and 268,000 cattle would allow the prevalence of BSE to be determined with 95 to 99 per cent accuracy, but otherwise refused to specify any specific target for the tests.
This could be because it is not clear how government testers will get access to these "downer" animals, which since December have not been accepted as human food and are not taken to

abattoirs. DeHaven said he hoped samples could be acquired on farms, at veterinary clinics and rendering plants.
But veterinary experts fear many farmers could quietly kill and bury their downers, rather than risk a test that, if positive, would mean the destruction of their herd.

Double checked

The international panel also recommended testing some apparently healthy animals at abattoirs. Switzerland implemented this policy to discourage farmers from sending suspect animals for slaughter before their symptoms become obvious. DeHaven said the US will test only 20,000 slaughtered animals.
Cattle will be tested with the fast tests now used in European abattoirs, with any positives double-checked by immunohistochemistry at the USDA lab at Ames, Iowa.
"We expect that there will be positive results on these screening tests, that's just the nature of the beast," said DeHaven, noting that these might be false positives.
But, in May 2003, US officials acknowledged in a report to the World Organization for Animal Health that some fast tests do not have this problem. The Swiss company Prionics says its western blot test has given no false positives in 18 million tests in Europe.

Two cows have been discovered with a form of BSE that looks very different from the usual kind, Italian scientists have reported.

It resembles one form of the human prion disease, sporadic CJD, raising the possibility that this human disease is acquired from cattle. Unlike vCJD, which is caused by eating BSE-infected beef, sporadic CJD is thought to occur spontaneously.

However, other scientists caution that many more than two cows will have to be found before it can be concluded that a new form of disease has been discovered. It may be that the two cows simply caught BSE via a different method of infection.

In transmissible spongiform encephalopathies (TSEs), of which BSE is one type, an insoluble form of a protein called a prion accumulates in the brain - with fatal consequences. In different TSEs, these accumulations look different.

In cows with BSE, the prion deposits lead to spongy holes, primarily in the brainstem and lower brain. But Salvatore Monaco, of the University of Verona, reports that two cows that tested positive for BSE at normal slaughter in an Italian abattoir in 2003 had completely different patterns of deposition.

Layered arrangement

Instead of the brainstem, there were prion deposits in higher areas of the brain, the thalamus and the olfactory bulb. The deposits were also arranged in layers called amyloid, which is found in vCJD, but rarely in cattle.
Furthermore, the prion protein was less glycosylated - modified by having sugars attached - than the usual BSE protein, and was lighter when tested by electrophoresis.
It could be the same BSE prion as usual, but deposited differently in the brain because it was inhaled rather than swallowed, Monaco suggests, as the prion was also found in olfactory tissue. However, it might also be a separate TSE.
If it is, it could be fairly widespread, as earlier studies found amyloid in one in 20 cows with BSE. Monaco notes similarities between this form of cattle TSE, which he calls Bovine

Amyloid Spongiform Encephalopathy (BASE), and one of the six types of sporadic CJD, M/V2, which has similar glycosylation and particle size.

Abattoir testing

The two cows with BASE were detected by a BSE test made by the Swiss company Prionics. Markus Moser, at Prionics, says that even if BASE is a distinct form of BSE, it is not likely to escape being detected by the tests used in European abattoirs.
This should mean it does not pose an added risk to people eating beef in places such as Europe, where older beef cattle must pass a BSE test. But if it is a second cattle TSE, it could arise spontaneously in places with no history of BSE infection. This would pose a potential risk, especially as those places allow cattle brain in food.
To prove BASE is a second TSE, Moser says the same pattern of chemical properties and deposition will have to be found in far more than two cattle. Then the prion must be injected into the brains of mice and shown to cause a different pattern of damage than the usual BSE prion. It must also maintain its distinctive properties when put back in cattle.
The strain is now being tested in mice, but the tests can take years. If it does have the same effect as M/V2 CJD prions in mice, it would raise the possibility that M/V2 CJD is not spontaneous after all, but could be caused by BASE in cows.
In 2001, French scientists were shocked when a strain of scrapie from French sheep looked identical to another form of human sporadic CJD in mouse tests. Scientists at the time said this meant an infectious cause for sporadic CJD could "no longer be ruled out", but further studies have yet to be concluded.
Journal reference: *Proceedings of the National Academy of Sciences (DOI: 10.1073/pnas.0305777101)*

US faces up to BSE; 10 January 2004

The US government has moved fast to try to save its $400 billion beef industry after a BSE-infected cow was discovered just before Christmas. But a quarter of Americans surveyed say they will eat less beef, and export markets remain closed amid charges that the US has not yet done enough to contain the disease.

Last week, US agriculture secretary Ann Veneman finally defied meat-industry opposition by banning cattle that cannot walk from being used in human food. In Europe, these "downers" are 10 times as likely as the rest of the herd to carry BSE. Veneman also banned the use of cattle brain and spinal cord in human food, including recovered meat slurries.

European scientists have long predicted that BSE would be found in the US (New Scientist, 5 July 2003, p 5), and the lack of safeguards until now means that some Americans may have been infected with variant CJD. How many will depend on how much infection there is in US cattle, and this will only be known if many more animals are tested. Last year the US tested just 20,000 of its 100 million cattle, enough to prove only that fewer than 13,000 animals are infected. US officials have not yet announced any plans to increase testing.

Meanwhile, downers and other potentially infected cattle will still end up in pig and poultry feed, which could spread infection back to cattle - by contamination of cattle feed, for example.

US bans comsumption of cripped cows; 31 December 2003

Crippled cattle are to be banned from human consumption, the US government announced on Tuesday. The move follows the discovery of the first case of BSE in an American cow - which was a crippled or "downer" cow.

The cow was confirmed as positive for BSE on 25 December, after it was slaughtered for food in Washington state earlier in the same month. Meat from the cow was recalled and its herd and offspring were quarantined.

The discovery confirms the longstanding warnings of European veterinary experts that BSE could be present in the US. But stringent controls, including banning crippled cattle from human food, have been resisted.

The US Department of Agriculture has been testing some 30,000 US cattle a year for BSE since 2001, targeting downers because European scientists found such cows were most likely to reveal the presence of BSE in a herd. A downer first revealed the presence of BSE in Canadian cattle in May 2003.

Some 20,000 downers are eaten yearly in the US. Canada and European countries have banned such cattle from human consumption. But moves to ban their consumption have been widely resisted by the powerful US meat industry. Prior to the new announcement by US agriculture secretary, Ann Veneman, the US National Cattlemen's Beef Association had told journalists it would continue to resist efforts to declare all downers unfit to eat.

However, following the move, the association said it would back the government's actions. The ban on downers will take immediate effect.

Cattle feed ban

US officials are stressing that the infected cow was born in Canada. But Canadian agriculture minister Bob Speller told a press conference in Winnipeg on Monday that it might still have contracted the infection from cattle remains in feed that came from the US. The two countries' beef industries have been closely linked for decades.
The cow was born four months before a ban on using cattle remains in cattle feed took effect in the US and Canada in 1997. US officials stress that, even if there was some infection in the US herd then, the feed ban would have kept it from spreading.
But critics, including Swiss scientists who reviewed Canada's similar BSE controls earlier this year, say infection could still have spread. Cattle remains are still permitted in feed for chickens and pigs - and the new measures announced by Veneman will not change this. When the European Union had similar rules, substantial BSE contamination still managed to enter cattle feed - leading European countries to ban cattle in all feed.
The new measures will, however, ban the use of "specified risk material" such as spinal cord, brain and eyes from cattle over 30 months of age from entering the human food supply.

VCJD death linked to blood transfusion; 17 December 2003

A British man has died from variant CJD after receiving a blood transfusion seven years earlier from a donor who also later died from the disease. The UK secretary of state for health announced the case in an emergency statement to parliament on Wednesday.

The case may be the first in the world where the human form of BSE - mad cow disease - has been transmitted via a blood transfusion. But John Reid told the House of Commons: "This is a possibility not a proven causal connection."

Both individuals might have acquired the devastating illness separately by eating BSE-infected meat, he says, but "the possibility of this being transfusion-related cannot be discounted".

The recipient of the blood transfusion had received the blood in 1996 from a donor with no sign of vCJD. But the donor developed VCJD symptoms and died in 1999. The recipient became ill six and a half years after the transfusion and died in autumn 2003.

Fifteen other people in the UK are now known to have received blood from donors who subsequently developed vCJD, according to the UK National Blood Service. These individuals are now being contacted and will be offered advice from expert counsellors.

No test

"There is as yet no blood test for vCJD, or for that matter BSE, let alone one that could detect the disease years before symptoms develop," said Reid. "So, there is no way yet of screening blood donations for the presence of the CJD group of diseases."
However, he noted that the UK Government had put measures in place from 1997 to try to reduce the risk of person-to-person transmission of vCJD via blood transfusions. First, blood stocks from donors who later developed vCJD were destroyed.
In July 1998, a programme to remove the white blood cells from blood destined for transfusion began. White blood cells were considered to be a possible source of infection. Finally, from the end of 1999, all plasma used for blood products has been sourced from the US rather than the UK.
However, 15 people have received blood from donors who subsequently developed vCJD. Five of these received the "leuco-depleted" blood - stripped of its white blood cells. But "many more patients of course will have received plasma products before plasma was sourced from the US," cautioned Reid.

Uncertain future

The ultimate extent of the UK epidemic is currently estimated to be between a few hundred and 7000. So far 143 people had died of the disease.
"**It is premature to conclude the epidemic has peaked" warns Reid, because the incubation period of vCJD is uncertain.**
The link between the recent death of the blood recipient and the donor was first reported to Reid's office on 9 December, while a diagnosis of vCJD for the recipient was still being confirmed.
Confirmation came on Friday 12 December and Reid was briefed by the UK's chief medical officer Liam Donaldson on Monday and Tuesday. The announcement was made at the "earliest opportunity", says Reid.

Protein locks out prion diseases; 4 October 2003

A chance discovery could lead to the development of a drug that blocks prion diseases such as variant CJD. However, it would only be useful when combined with mass screening to identify infected people who have not yet developed any symptoms.

Adriano Aguzzi's team at the Institute of Neuropathology in Zurich, Switzerland, discovered the effect by accident. The researchers engineered mice to produce a protein that would stick to the scrapie prion, so the team could retrieve the prion protein for testing. The engineered mice turned out to take twice as long as normal to develop the disease.

Because the protein is soluble, it is ideal for use as a drug. Aguzzi now hopes to mass-produce the protein so he can test it on macaque monkeys that have been exposed to BSE.

If it works, Aguzzi told the European Life Science Organisation conference in Dresden last week, his team will test it on people infected with vCJD.

Other drugs are also being developed (New Scientist print edition, 8 March 2003), and some are being tested on people. Last week doctors announced that injecting a substance called pentosan polysulphate into the brain of a teenager in Northern Ireland who has vCJD seems to have slowed the disease's progress.

But fears that thousands of people in the UK are incubating the disease after eating infected beef are receding. There have been only 136 cases so far, and the latest estimate is that only another 40 people will develop vCJD.

Bovine spongiform encephalopathy (atypical case)in Japan; 29 September 2003

Description of affected population; a Holstein bullock aged twenty- three months slaughtered at the abattoir on 29 September 2003.
Diagnosis;
-The brain sample from the bullock tested positive to the ELISA-based BSE screening test, and was sent to the national Institute of Infectious Disease for confirmation and was subjected to Western blot analysis, histopathological examination and immunohistochemical examination. Based on these results, the case was concluded as an atypical BSE on 6 October 2003.
-Result of Western blot analysis; PrPsc (scrapie-associated prion protein) was detected, the pattern of glycoform and relative protease resistance of PrPsc were different from what is known for BSE. Results of histopathological examination and immunohistochemical examination were negative.

Volcanic pool enzyme kills prions; 29 July 2003

A new disinfectant, based on enzymes collected from a volcanic pool, is showing promise in destroying the mutated prion proteins that cause vCJD, the human form of BSE.

These prions are notoriously difficult to break down, and because an unknown number of people in the UK have vCJD, there is a theoretical risk that surgical instruments could transmit the disease. The disinfectant could also be effective against the prions that cause sporadic CJD, which occurs spontaneously and has been spread surgically.

Standard decontamination procedures, such as detergents, UV inactivation and high-pressure boiling at 137 °C, have proven unsuccessful at destroying all the prions on contaminated equipment.

But now researchers at the UK's Centre for Applied Microbiology and Research (CAMR) in Porton Down and biotechnology company Genencor believe they have developed a prion eradication agent.

"Essentially it's a protease enzyme, which is active at high alkalinity - pH 12 to 14 - and a temperature of between 60 and 80 °C," says CAMR's Phil Luton. "It requires an incubation period of less than one hour under the enzymes' optimal conditions to degrade the prions."

Bond breaker

The researchers tested a variety of different enzymes, before selecting the most effective. This was a genetically modified version of a naturally occurring, thermostable enzyme found in volcanic pools and was produced by Genencor.
"The enzyme appears to be very promising from the preliminary research findings, although it will need to undergo further tests," says UK-based prion expert John Stephenson. "It works by chemically breaking down the bonds between amino-acids in the protein."
He told New Scientist: "We don't know why prions are so highly stable, but they are extremely hard to destroy. Indeed, one standard method of decontamination - soaking in fomaldehyde - actually stabilises the prions."
Stephenson notes that several other research groups are exploring similar ways of decontaminating surgical instruments.

Tonsil trouble

Safety procedures issued by the UK Department of Health (DoH) in 1999 require surgical instruments used on patients with suspected vCJD to be quarantined until diagnosis is confirmed or discounted.
"The surgery of highest risk is brain or eye surgery, although tonsillectomy also carries risk," says Stephenson. "**Studies have shown that tonsils removed from patients with vCJD have quite high levels of infected prions, although infection has never been found in tonsils from those with the sporadic CJD."**
In January 2001, the DoH ordered the use of disposable tonsillectomy kits, but this was reversed in December 2001 after the kits caused a rise in surgical complications.
Since 1990, 132 people have died of vCJD in the UK, with 659 succumbing to sporadic CJD. **The number of people infected with vCJD remains unknown and there is no diagnostic test. Epidemiological data suggest the epidemic is still growing, with a final death toll as high as several thousand.**

BSE and the US; 21 June 2003

Debora MacKenzie makes claims about US efforts to prevent bovine spongiform encephalopathy, despite the fact that no evidence of BSE has ever been found in the US (31 May, p 6).

Since 1989, the US Department of Agriculture's Animal and Plant Health Inspection Service has put in place numerous safeguards to prevent BSE from entering the US. These safeguards include stringent import requirements and BSE surveillance levels that far exceed international standards.

In the fiscal year 2002, the US tested more than 19,900 "high-risk" cattle. Experience from Europe confirms that testing non-ambulatory cattle and cattle that show signs of central nervous distress is the best way to discover whether BSE is present. At our current surveillance level, we would detect BSE at a rate of one case per million head of cattle.

In addition to targeted surveillance efforts, the US prohibits the use of most mammalian protein in the ...

Britain could resume eating older cattle; 18 June 2003

Britain could drop its expensive ban on eating cattle over 30 months old without significantly raising the risk of vCJD in people, according to a newly-published epidemiological analysis.

Currently, all older British cattle are incinerated to stop BSE entering the human food chain. In contrast, in the rest of Europe, cattle are tested for BSE at slaughter. The epidemiologists calculate adopting this approach in Britain would most probably cause no extra vCJD deaths over the next 60 years.

The "over 30 months" (OTM) rule was introduced in 1996, when scientists realised that eating BSE-infected cattle caused the lethal human disease vCJD. Almost no cattle infected with BSE get sick before this age, so the infectious agent is thought to be at low levels in young cattle - making them safe to eat.

The OTM rule has sent more than six million cattle to the incinerator. The British government compensates farmers and disposes of the carcasses, at a cost of £420 million per year.

Falling incidence

The incidence of BSE in British cattle has fallen steadily since 1996, leading the UK Food Standards Agency to start reviewing the OTM rule in 2002.
In March 2003, a "core stakeholder group" of scientists, consumers and farmers recommended that the OTM rule be dropped, either completely or for those cattle born after 1996.
This recommendation was based on the best estimate calculated by "independent experts" that switching to testing at slaughter would lead to only 0.04 additional deaths over the next 60 years. Using the most pessimistic assumptions, the figure rose to 1.6 deaths. The stakeholder group also noted that dropping the OTM rule would reduce costs by 93 per cent.
The independent experts have now been revealed as Neil Ferguson and Christl Donnelly of Imperial College London. They have published the details of their calculations in the *Proceedings of the Royal Society B*, meaning others can submit comments on the work to the Food Standards Agency review, which closes in early July. After that the FSA will make its proposals to government.

Test sensitivity

Ferguson and Donnelly calculated the prevalence of BSE now in British OTM cattle, and assessed how much could be kept out of the food chain with testing.
One key uncertainty is how many cases of vCJD results from a given amount of BSE exposure. Other uncertainties are the sensitivity of the BSE tests and the likelihood that cattle incubating BSE will be sent for slaughter just before they show obvious symptoms.
Even using pessimistic assumptions for these factors, the increase in deaths from vCJD over the next 60 years remained below one. Only factoring in an unlikely underestimate of BSE prevalence pushed the estimated deaths above one, to 1.6.
Journal reference: *Proceedings of the Royal Society B* (DOI 10.1098/rspb.2003.

BSE crosses the Atlantic; 31 May 2003

The US and Canada were warned that some of their cattle might have mad cow disease. But neither country has been testing enough animals to rule this out

The discovery of a BSE-infected cow in Canada confirms warnings from European scientists three years ago that cattle in North America could be infected. And if Canadian cattle are infected it is likely that the disease is also present in the US.

The US and Canada test so few animals that low levels of BSE infection would not be detected. Indeed, the number Canada tests would be unlikely to reveal a level of infection any lower than what the UK now has. Other countries have found many more cases after increasing testing when the first infected cattle were reported.

What's more, neither country has taken any of the measures needed to prevent people being infected by meat from diseased cattle (see "What to do now"). Even if people are eating infected meat, there are unlikely to be many cases of vCJD, the human disease linked to BSE. In the ...

Mad cow quarantine in Canada; 23 May 2003

Canadian officials quarantined two more cattle herds on Thursday, as investigators scrambled to find out how a single cow contracted mad cow disease, and if any other animals in North America are infected.

It was announced on Tuesday that a cow slaughtered in Alberta in January had tested positive for bovine spongiform encephalopathy (BSE). The same disease led to the slaughter of millions of British cattle in the 1990s. The human form, vCJD, has killed more than 120 people so far.

The newly quarantined herds brought the total number to nine, said Claude Lavigne, an official with the Canadian Food Inspection Agency. It is still not clear where the infected cow originated, but it seems increasingly likely to have been born in Canada. The only other case of BSE in North America was in a cow imported into Canada from the UK in 1993.

Lavigne said the cow might have had as many as five calves that could have been eaten by humans.

Eating steak

Officials and the industry insist this is an isolated case and that testing procedures already in place mean beef is probably safe. Prime Minister Jean Chretien invited photographers to take his picture eating a steak on Tuesday.
"Canadian beef is safe to eat today, it was safe to eat yesterday, and it will be safe to eat tomorrow," said Cindy McCreath, a spokeswoman for the Canadian Cattlemen's Association.
But the US and most of Canada's other trading partners immediately banned imports of Canadian beef, a move that could cripple the billion dollar industry. Canada is the third-largest beef exporter in the world.

Rapid transmission

In 1997 Canada and the US banned the feeding of protein from cattle and sheep back to cattle. This practice is thought to have caused the rapid and widespread transmission of the disease in the UK. Canada and the US also test several thousand animals a year for BSE.
Critics claim holes in the safeguards and monitoring efforts could have allowed the disease to spread. But US regulators point to a study conducted in 2001 by the Harvard Center for Risk Assessment which concluded that the US is unlikely to develop any cases of BSE, and if it does, that measures already in place should contain it.
If correct, the conclusion would hold true for Canada too, since the two countries share similar food safety regulations.

Canada finds case of mad cow disease; 21 May 2003

Canada has announced its first case of "mad cow disease" for a decade, prompting an immediate ban by the US on Canadian beef.

But Canadian government officials and cattle farmers are insisting the meat supply is safe, despite the revelation of the case of bovine spongiform encephalopathy (BSE) on Tuesday in a slaughtered cow. The herd of 150 cattle in Alberta, from which the infected cow came, has now been quarantined and will be destroyed and tested for the deadly disease.

"We remain confident in our beef and cattle industry," said Shirley McClellan, Minister of Agriculture, Food and Rural Development for Alberta - home to nearly half of Canada's cattle.

The case is the first in Canada since a cow imported from Britain was diagnosed with BSE in 1993. That cow contracted the infection in Britain. Canada has banned imports from BSE-affected countries, and in 1997, it prohibited adding protein derived from ruminants - including cows and sheep - to feed destined for cattle. It has insisted that no cow has ever acquired BSE in Canada.

Canadian officials said their first priority is to determine how the cow contracted the disease. Lyle Vanclief, Federal Agriculture and Agri-Food Minister, said it was not immediately apparent whether the cow had been imported or not.

Unfit for consumption

The infected cow was slaughtered in January, but was rejected as unfit for human consumption by a government inspector who suspected the animal suffered from pneumonia.

Test results were not available until 18 May, because the tests had been assigned as low priority given the animal displayed no symptoms of BSE. Canadian officials immediately quarantined the Alberta farm where the cow originated. The BSE World Reference Laboratory at Weybridge, UK confirmed the presence of BSE on Tuesday.
The US "is placing Canada under its BSE restriction guidelines and will not accept any ruminants or ruminant products from Canada pending further investigation," said Ann Veneman, US Agriculture Secretary. The US has never reported a case of BSE.
But Veneman and Canadian officials are emphasising the case is an isolated one, and say the risk to human health and other animals is low. The task now, they say, is to trace the origin of the infected cow, in case any of its herd mates were also infected.

Maximum risk

According to data Canada provided to the European Commission in 2000, no cattle have been imported into Canada from any countries known to have BSE since 1990, before the infected cow was born.
In their assessment, Commission scientists concluded that it "cannot be excluded" that native Canadian cattle are infected with BSE. This is because eleven cattle carrying BSE were probably imported from Britain before 1990, and they would have been recycled into cattle feed in Canada with no precautions against BSE. The Commission reached a similar conclusion about the US.

Any infection this caused in Canadian cows would then have been recycled, and multiplied, as those cows also entered cattle feed before 1997. The Commission scientists calculated that this would have posed a maximum risk between 1993 and 1998 - the infected Alberta cow was born in 1995. Most cattle are thought to be infected by BSE in feed when they are young.

Patient benefits from controversial vCJD drug; 12 May 2003

An experimental drug given to a UK teenager with the human form of mad cow disease appears to have stopped the fatal disease in its tracks. The controversial treatment may even have improved 18-year-old Jonathan Simms' condition, according to his father.

Pentosan polysulphate had never been used in humans for treating vCJD before being injected directly into Jonathan's brain. The treatment began in January 2003 after the family won a High Court ruling against the UK's National Health Service.

The NHS had refused to allow doctors to carry out the procedure with an untested drug. But the court ruled the treatment acceptable, as without treatment Jonathan was certain to die.

His father, Don Simms told a BBC documentary: "I can categorically state that Jonathan has not got any worse. He in actual fact shows signs of improvement. We are not hailing it as a total success, but from what we have seen so far we are much encouraged."

Stephen Dealler, a medical microbiologist the Royal Lancaster Infirmary and an expert on prion disease, said he felt "very enthusiastic" about the treatment. "I was really taken aback by this," he told **New Scientist**. "At this point there have been significant physiological improvements and no side effects whatever."

Extended incubation

Pentosan polysulphate is a commonly used oral medicine for cystitis and bladder pain. It has shown positive effects in mice with scrapie - a disease similar to CJD. Japanese and UK researchers showed that brain and abdominal injections given shortly after infection significantly extended the incubation period of the disease.
Few side effects were seen in mice but as the drug had never been tested as a vCJD treatment in humans, scientists were uncertain about the what dosage would have an effect without causing an adverse reaction or even aggravating the devastating illness.
Jonathan, from Northern Ireland, was given 12 infusions each at a gradually increased dose. "His pulse rate is down to more normal levels, the salivating associated with vCJD has dramatically decreased and he looks much better. He has gained weight and his face has returned to a normal colour," Don Simms told the website *ic Northern Ireland*.

Mutant protein

Dealler says test tube experiments have shown that pentosan could theoretically tackle vCJD in two ways. Firstly, minute quantities of the drug can build up in the brain and to stop the production of the mutant prion proteins thought to cause vCJD.
Secondly, it could cut the activity of inflammatory molecules called interleukins, whose production is stimulated by the misshapen prions. The interleukins attack brain cells, which ultimately leads to their death.

Previous promising vCJD treatments have in the end failed. But Dealler is optimistic about pentosan because of the lack of side-effects. Quinacrine drastically improved the condition of a UK sufferer in 2001. But Rachel Forber was taken off the drug after suffering liver failure. She died of causes unrelated to this damage within months.

Predicted deaths from vCJD slashed; 26 February 2003

The worst case scenario for the deaths caused by vCJD, the human form of mad cow disease, has been revised downwards from 50,000 to 7000 by a new analysis.

In 1997, the UK research group predicted that up to 10 million people could die from the devastating disease. In 2002, the figure dropped to 50,000, based on data up to 2000. Now researchers at Imperial College, London say the likely upper limit of deaths has fallen to 7000.

Azra Ghani and colleagues used epidemiological data to model vCJD cases and deaths. Their best estimate now is that 80 more deaths will occur by 2080 - 122 have already died in the UK. However, there is still a lot of uncertainty, says Ghani.

"**The large numbers [predicted] are now looking very unlikely - because you would have had to have seen an awful lot of cases by now," Ghani told "But obviously 80 deaths is still 80 deaths."**

She notes that the upper limit of 7000 was based on an analysis of data up to the end of 2001. The group has since added the data for 2002 to their analysis and submitted the results for publication. "It does look like it will be even lower [than 7,000], as we have had two years in a row now where numbers look to be declining."

Confidence limit

The upper limit of 7000 deaths by 2080 represents the upper 95 per cent confidence limit. The equivalent lower confidence limit was 10 deaths.
Short term projections by the group also show that a dramatic increase in cases is unlikely. In the next two years, they predict 30 deaths, with upper and lower limits of 10 and 80. And in the next five years, they predict 70 deaths, with upper and lower limits of 10 and 200.
The group only looked at cases and deaths from people thought to have become infected with vCJD by eating infected meat. Their study does not account for secondary transmission of the illness, which could theoretically occur from human to human via infected surgical instruments or blood transfusions.
Ghani said a scarcity of data made it "very difficult" to say what effect this might have on a possible epidemic. Other researchers are currently examining people's tonsils to try to determine directly the level of infection in the population.
Another factor not included in the model is the possibility that some people are more susceptible to vCJD than others. All the cases so far have occurred in people with particular versions of genes related to the prion protein that is the key to the disease. These people make up 40 per cent of the population
The remaining 60 per cent "could either be less susceptible or have a longer incubation", says Ghani. This could at worst double the number of deaths, she says. **But if incubation periods are longer, perhaps 25 years, then people are more likely to die of other causes first.**
Journal reference: *Proceedings of the Royal Society B.* (DOI 10.1098/rspb.2002.2313)

BSE may cause more CJD cases than thought; 28 November 2002

The eating of BSE-infected meat might cause classical CJD in people, as well as variant CJD, **a new mouse study suggests.**

Classical CJD, also called sporadic CJD (sCJD), is generally believed to develop spontaneously and existed before the BSE epidemic in British cattle, while variant CJD (vCJD) is thought to be the human form of mad cow disease.

There has been a recent rise in cases of sCJD, in the UK in particular, but it was thought this was due to better surveillance and diagnosis. But the surprising new finding adds weight to suggestions that the rise is in fact linked to the BSE epidemic.

The new work involved injecting the BSE infectious agent - a misfolded prion protein - into the brains of mice. The mice had been genetically modified to act as human models of infection and to be susceptible to CJD.

As expected, some of these mice developed symptoms and a molecular subtype of prion protein misfolding associated with vCJD. But others developed a sub-type associated with the most common of three strains of sCJD previously identified in people.

"This finding has important potential implications," the team led by John Collinge at the MRC Prion Unit in London, UK, writes in the *EMBO Journal*. "It raises the possibility that some humans infected with BSE prions may develop a clinical disease indistinguishable from classical CJD."

Models predicting the future extent of the human epidemic associated with eating BSE-infected meat are based on the observation of vCJD in the population. But if BSE prions can also cause sCJD, these models will underestimate the ultimate human death toll. To date, 117 people have died in the UK from vCJD.

Steady rise

More sCJD deaths are recorded annually than vCJD deaths. In 2001, 53 people in Britain died from sCJD and 20 from vCJD. This compares to 35 from sCJD and three from vCJD in 1995, when the variant form was first identified.

The steady rise in the recorded incidence of sCJD has been put down to better detection. "**What we're speculating is that a proportion of that rise, not all of it but a proportion, is due to BSE," Collinge says.**

In Switzerland, which had the highest incidence of cattle BSE in continental Europe between 1990 and 2002, there has been a doubling in the number of sCJD cases in that period.

But establishing how many, if any, cases of sCJD in people resulted from BSE infection will be very difficult, Collinge says. The two diseases cause similar symptoms, and the molecular type of sCJD seen in the BSE-injected mice is indistinguishable from the human type identified before the first cases of mad cow disease.

This is not the first study to link eating meat from an animal infected with a prion disease to sCJD. In March 2001, a French team found that one strain of scrapie - a prion disease present in sheep for centuries - can cause the same brain damage in mice as sCJD. This suggested that some cases of sCJD in people might be down to eating scrapie-infected sheep.

Journal reference: *The European Molecular Biology Organization Journal* (vol 21, p 6358)

Surgery patients exposed to CJD risk; 30 October 2002

Twenty-four surgery patients treated at a hospital in the northeast of England are being told that they could have been infected with Creutzfeldt-Jakob disease (CJD).

The risk, thought to be very small, has arisen because instruments used to operate on a patient with sporadic CJD were re-used. The UK Department of Health (DoH) described the incident as "appalling" and said the hospital had apparently "failed to prevent avoidable and unnecessary exposure". The government's chief medical officer has now ordered an inquiry.

Instruments used to conduct a brain biopsy on a female patient at Middlesbrough General Hospital were later used on other patients. But two weeks after the biopsy on 29 July, the woman was confirmed to be infected with the deadly brain disease. Only then were the instruments removed from use. There are five recorded cases of CJD transmission following neurosurgical operations.

The hospital maintains that prior to the biopsy, she was examined by five neurologists, none of whom suspected she had the disease. However, Paul Lawler, medical director of the NHS trust that runs the hospital has admitted that "in hindsight" it may have been better to have removed the instruments immediately, rather than waiting until the CJD diagnosis had been confirmed.

Mutated protein

Safety procedures issued by DoH in 1999 require surgical instruments used on patients with suspected CJD to be quarantined until diagnosis is confirmed or discounted. This is because the mutated prion protein though to be responsible for CJD may survive standard decontamination techniques.
The precautions were introduced because an unknown number of people in the UK are carrying variant CJD, following the BSE fiasco of the 1980s and 1990s. But the incident in Middlesbrough will raise serious questions about how well the precautions are being complied with.
"Patients are potentially at risk from people who are carrying vCJD, but don't know it. It's an unknown quantity," says a spokesperson from the National Prion Clinic at St Mary's Hospital, London.
Sporadic CJD is a prion disease that occurs randomly in the population and has no known cause. The other form of the disease, vCJD, is believed to be transmitted by eating meat from BSE-infected cattle. Both forms lead to brain deterioration and eventual death, and there is no treatment or cure.

Since 1990, 577 people have died of sporadic CJD in the UK, with 117 succumbing to vCJD. The number of people infected with vCJD remains unknown and recent data shows the epidemic is still growing. The maximum estimates of the ultimate extent of the epidemic suggest vCJD will afflict thousands of people.

Loop- shaped saw cuts BSE contamination risk; 19 October 2002

A novel power saw that can strip the spines out of cattle and sheep carcasses should further cut the risk of infecting people with the agent that causes the fatal brain disease vCJD.

Spinal cord from cattle has been banned from food in Britain since 1996, when the link between mad cow disease (BSE) and vCJD was established. If an animal is incubating BSE, its spinal cord could harbour the defective prion protein that causes the disease in both animals and humans. Abattoirs normally use a bandsaw to split the carcasses of cattle in half. But if the saw cuts through the spine, material from the spinal cord is likely to contaminate other parts of the carcass. The new saw cuts out the spine before the carcass is split.

Is an epidemic of CJD going undetected?; 11 May 2002

The brain-wasting disease CJD may be much more common in young people than anyone had suspected.
Two young men from Michigan who died from the disease last autumn were only diagnosed because a specialist insisted on tests that wouldn't normally be done. He is now saying that when anyone, no matter how young, dies with suspicious neurological symptoms, there should be a post-mortem to check for CJD.
The men, aged 26 and 28, were brought to the University of Michigan hospital in Ann Arbor last summer with rapidly worsening neurological symptoms. One had severe epilepsy, the other had symptoms of dementia. Norman Foster, the attending neurologist and a CJD specialist, says neither man's symptoms fitted all the official criteria for the disease.
Normally they might have been diagnosed as suffering from brain inflammations of unknown cause, he says. "But we pushed to get a biopsy." It revealed that each had a different form of sporadic CJD.
The pattern of brain lesions and an analysis of the prion proteins that caused the disease showed it was not vCJD, the human form of BSE. Nor did it look like the chronic wasting

disease of deer that occurs in neighbouring Wisconsin, which some fear might also be transmissible to humans.
Foster warns that if infection from deer led to an upsurge in CJD, it would go undetected because the disease is rarely tested for in young people. He thinks anyone who dies with dementia should be autopsied, and brain biopsies should be taken from young people who might have CJD, even if they do not have all the usual symptoms. He and other CJD specialists will meet in October to plan a surveillance programme for Michigan.

Search for BSE in muscle meat draws blank; 27 March 2002

Tests by government scientists in France have allayed renewed fears that eating beef can cause variant Creutzfeldt Jakob Disease, the human form of BSE. The fears were heightened on 18 March when scientists unexpectedly reported finding traces of infective material in the muscles of mice.
The findings carried extra weight because they came from the lab of Stanley Prusiner, the scientist at the University of California in San Francisco who won a Nobel Prize for discovering mutated "prions". These are the defective proteins believed to cause brain-wasting diseases like BSE, vCJD and scrapie.
Prions are known to collect in brains, spinal cords, spleens and other lymphoid tissue and these parts have long been banned for human consumption. Muscle tissue in meat was assumed to be free of prion contamination and safe to eat.
But Prusiner's team found prions in the hind leg muscles of mice whose brains had been injected with BSE-like prions. He reported his results in Proceedings of the National Academy of Sciences.

Complete blank

The new twist in the story came on Wednesday, **when French government scientists announced that they had drawn a complete blank when they looked for prions in muscles from several BSE or scrapie-infected animals including mice, sheep, goats and cows.**
"The tests proved negative in the search for pathological prions in the set of samples, including those taken from the hind limb muscles," says the AFSSA, France's food safety watchdog.
Reassuringly, tests on peripheral nerves and lymphoid tissue in the cow also came up negative, suggesting that meat will be safe even if it contains this type of tissue. "These observations are consistent with the findings made to date concerning the distribution of the infectivity linked to BSE in cows," they say.
The researchers detected prions with two tests. The first, an "ELISA" test, detects antibodies made by animals against prions. The second "Western blotting" test isolates and identifies fragments of the prion protein itself. The scientists will discuss their results in more detail at a meeting of France's top BSE specialists on 11 April.
Low level infectivity
The significance of Prusiner's results has also been questioned in Britain. Peter Smith, chairman of the government's Spongiform Encephalopathy Advisory Committee, told New Scientist that a much more significant experiment had been under way for five years in Britain, and had yet to give any cause for concern.
Instead of using a surrogate animal like the mouse, this experiment is focusing on which cattle tissues can transmit BSE to other cattle. Scientists at the Veterinary Laboratories Agency in Weybridge, Surrey, injected the brains of live calves with liquidised tissue from various parts of the bodies of BSE-infected cows.
"None injected with muscle have gone down yet," says Smith. "But if you inject BSE-infected brain tissue, the calves come down with BSE in about two years, as expected," he says.

"If there is infectivity in muscle, it must be at a much lower level than in other tissue, particularly that from the brain or central nervous system," says Smith. You can never prove a negative, he adds, **but the results so far have been reassuring**.

Britain: Report highlights BSE danger from infected sheep; 21 January 2002

The risk to humans developing variant Creutzfeldt-Jakob Disease (vCJD) could be far greater if the brain-wasting disease Bovine Spongiform Encephalopathy (BSE) has entered the sheep population. This was the conclusion of a study published in the British science magazine *Nature* on January 10.

The study was carried out by researchers working in the infectious diseases department of Imperial College London led by Professor Neil Ferguson.

BSE in cattle, also known as “mad cow disease”, is believed to have been spread by the practice of feeding cows the rendered remains of slaughtered cattle and other livestock. Until legislation banned the practice, sheep were also fed the same material.

Since it began in the late 1980s, the BSE epidemic has infected nearly 180,000 cattle. At its height in 1992 over 36,000 cattle had the disease. Numbers have now declined with around 700 cases last year.

Variant Creutzfeldt-Jakob Disease—the human form of BSE—is transmitted by eating infected meat or other animal products. Since 1995, 104 mainly young people have died of the disease, with nine more people currently diagnosed as suffering from this terminal and incurable condition. The eventual number of people who could be affected is still an unknown, because of the extremely long incubation period for the disease. There also remains the possibility of a second wave of infection via human-to-human transmission as a result of surgical procedures. Since the infective agent, the BSE prion, is extremely difficult to destroy, the usual sterilisation methods used on surgical instruments do not eradicate it.

The researchers at Imperial College considered three possible scenarios if BSE has passed into the national sheep flock. The worst possible case considered the effect of BSE spreading both within and between sheep flocks. The study’s median scenario projected the spread only within a flock, while the best-case scenario investigated what would happen if it spread neither between nor within flocks. Sophisticated mathematical models were devised to predict the possible effects on the human population.

In the worst case, the study predicts that 150,000 people could die as a result of eating infected sheep meat. This figure is three times higher than the worse case scenario of human deaths from vCJD contracted from eating contaminated beef.

It has not yet been shown whether sheep have in fact contracted the disease. The report is based on the assumption that BSE has passed from cattle to sheep and has been spreading from sheep to sheep. Many scientists think that such a cross over from cattle, and its subsequent spread within sheep, is a possibility. Professor Neil Ferguson said, “In some ways I’d be surprised if BSE wasn’t found in sheep.”

One difficulty detecting BSE in sheep is that sheep are also subject to a brain-wasting disease known as scrapie. This has been in the sheep population for 200 years and is considered harmless to humans. Currently there is no test to distinguish between BSE and scrapie in sheep.

Studies have shown that BSE in sheep behaves differently to the disease in cattle. It infects a wider range of sheep tissues at an earlier age. There are fears that BSE in sheep could mimic scrapie, which passes easily by horizontal infection from sheep to sheep.

Under current legislation, the ban on sheep offal is not as extensive as that on cattle offal, some of the most infective material. With sheep under 12 months old, only the spleen has to

be removed before the carcass can enter the human food chain. For sheep older than one year, the skull, brain, eyes, tonsils and spinal chord are banned, but not the lymph nodes or intestines (as in cattle).

Professor Ferguson said, “The current risk from sheep could be greater than that from cattle, due to the more intensive controls in place to protect human health from exposure to infected cattle, as compared with sheep.”

In a newspaper article in August last year, former government advisor Dr Richard Kimberlin warned of the potential danger from BSE-infected sheep: “We now know that several tissues from BSE-infected sheep, including lymph nodes, pose a greater risk than the same tissues from infected cattle”.

The Imperial College team says that banning all internal sheep organs from the human food chain would reduce the health risk by 90 percent.

Frances Hall, secretary of the Human BSE Foundation, said, “If it is in sheep, people could have been eating contaminated meat for years.” Frances, whose son Peter died from vCJD in 1996, added, “It’s very sad to think more families might be having to go through the same nightmare we’ve gone through needlessly”.

The Department for the Environment, Food and Rural Affairs (Defra) has begun a programme to screen sheep. Professor Tim Lang of Thames Valley University, who had written widely on food safety, is concerned about the suitability of Defra to conduct such a study. “One of the many things this sorry saga has taught us is that we couldn’t trust public health controls to be run by a ministry in charge of production.”

A previous government study set up to estimate BSE infection in sheep brains had to be abandoned last year. It was discovered that poor laboratory techniques meant the samples being studied were possibly either cow brains or sheep brains that had been contaminated with cow brains.

Following the BSE epidemic in Britain, the Labour government set up the Food Standards Agency (FSA). In October last year it issued an update report on the risk of BSE in sheep that stated, “If BSE were found to be present in sheep, the current SRM [Specified Risk Material, i.e. offal, spinal cord, etc.] controls would not be adequate to eliminate the risk of infected sheep meat form entering the food chain. It has been shown that it is impossible to remove all infectivity from a sheep.”

The FSA, which had commissioned the Imperial College report, issued an equivocal and defensive statement in response. It read, “We do not know whether BSE entered the sheep flock in the past and, if it did, whether it is in sheep today. Given this uncertainty, the agency has been proactive in examining whether further precautionary measures may be appropriate in addition to those currently in place.”

Although now at a much reduced level, the risk of BSE from cattle has not yet been eliminated. The FSA announced last week that the meat of a calf born to a BSE infected cow had entered the human food chain. The incident is reported to have occurred in Wales last November.

According to reports, a farmer had sold the calf to an abattoir. Normally, offspring of BSE-infected cattle would be culled and the carcass destroyed. Among the measures to combat BSE, a cattle “passport” system has been introduced, and this should have prevented the calf from being sent to the abattoir and its meat sold.

The FSA said a backlog on culling infected animals had built up because of the demands placed on the veterinary service by the foot and mouth epidemic, which was only declared officially over on January 15. Pressure on vets and other officials had led to a build up of suspect animals.

Defra published a report on November 2001 noting the continuing danger from BSE. “Despite the measures taken to control the current outbreak of foot-and-mouth disease in the UK, the

controls for restricting, slaughtering and testing BSE suspects continue albeit at a lower level due to the need to redeploy resources," the report said.
The FSA has called on the government to tighten up procedures to ensure that the offspring of BSE-infected cattle do not enter the food chain. Debby Reynolds veterinary director of the FSA said, "This is a regrettable incident" adding "We want to see the cull of offspring of BSE animals backlog cleared as a priority."

BSE: it´s not over yet; 12 January 2002

Thousands more people could die from the human equivalent of mad cow disease if Britain's sheep turn out to be infected with the disease, epidemiologists warn this week. They say that sheep offal, and sheep older than six months, should be banned from human consumption to ensure that any such infection cannot pass to people in the future.

But British authorities have still not managed to establish whether sheep are carrying BSE. Until they do, any further restrictions on an already stricken meat industry seem unlikely.

Sheep can catch BSE by eating infected feed, and they carry the infection in more of the edible tissues than cattle do. **This makes sheep potentially much riskier for human consumers. Worse, while cattle get BSE almost exclusively from feed, sheep can spread such prion diseases to each other, by nibbling on afterbirths for instance**. That means BSE could still be spreading among sheep ...

vCJD deaths will rise if UK sheep have BSE; 9 January 2002

Thousands more people could die from the human equivalent of mad cow disease if Britain's sheep turn out to be infected with the disease, epidemiologists have warned.

They say that sheep offal, and sheep older than six months, should be banned from human consumption to try to prevent any such infection reaching people in the future.

But British authorities have still not managed to establish whether sheep are carrying BSE. Until they do, any further restrictions on an already stricken meat industry seem unlikely.

Sheep can catch BSE by eating infected feed, and they carry the infection in more of the edible tissues than cattle do. This makes sheep potentially much riskier for human consumers.

Worse, while cattle get BSE almost exclusively from feed, sheep can spread such prion diseases to each other, by nibbling on afterbirths for instance. **That means BSE could still be spreading among sheep despite controls on feed.**

Look-a-like

British sheep are known to have eaten infected feed, but no one knows whether they actually got BSE because its symptoms look exactly like those of scrapie, an existing disease of sheep thought not to affect humans.
Government efforts to detect BSE in sheep that apparently died of scrapie were ignominiously abandoned in 2001, when the sheep brains being tested were found to have been contaminated with cattle tissue.

So Neil Ferguson and a team at Imperial College, London, modelled what would happen if BSE did infect British sheep. So far, 180 sheep that showed symptoms of scrapie have been tested for BSE in Britain, and all were BSE-free.
This means that the prevalence of BSE in sheep is not likely to be more than two per cent that of scrapie, which is endemic, the researchers at Imperial calculated. But this would still mean that sheep are now the main source of human exposure to BSE in Britain.

Food supply

If some sheep do have BSE, the degree of risk to humans depends on how readily it spreads. If it is not infectious enough in sheep to pass from flock to flock, the team predicts that the numbers of infected carcasses entering the human food supply should now be falling.
But if BSE spreads between flocks, the problems would just be beginning. The team's model shows that the number of infected sheep entering the British food supply would start rising steeply. This explosion of infected mutton would start about now, and increase fivefold by 2020.
Ferguson and his colleagues estimate that, at worst, 50,000 to 100,000 people could die by 2080 of vCJD acquired from cattle. If BSE is in sheep, and spreading between flocks, the maximum number of deaths could rise to 150,000. Yet the team found that nearly all these extra cases would be prevented if more precautions are taken with slaughtered sheep now.
Journal reference: *Nature* (DOI: 10.1038/nature709)

Bungled BSE experiments due to refrigerator mix-up; 30 November 2001

Bungled experiments to find out if sheep catch BSE probably went wrong because samples of cow and sheep brain got mixed up in a refrigerator. **That is the main conclusion from two independent audits to explain how the crucial experiment went so disastrously wrong.**

The audits blame the Institute for Animal Health's laboratory in Edinburgh for the mix-up. But the IAH's director, Chris Bostock, says the conclusions are unfair.

Everyone agrees the experiment is dead. "This is clearly an experiment that's flawed and uninterpretable," said Elliott Morley, The UK's minister for animal health.

Determining whether sheep can catch BSE is critical in understanding the epidemic and its effect on public health. No parallel experiments were run, meaning research to find if BSE was in the UK sheep flock in the early 1990s will have to be abandoned. Scientists are now focusing on whether today's flock is infected - the 400 animals examined so far have tested negative.

Bovine or ovine?

In 1997, IAH researchers injected what they believed to be sheep brains into mice, to see if the animals would subsequently develop BSE. The brains were originally collected in 1990 to 1992 from sheep that died of scrapie, the sheep equivalent of BSE. The researchers hoped to find out from the mice experiments whether, in some cases, the scrapie was in fact BSE.
But in October, just before the researchers presented their results, an independent analysis by the Laboratory of the Government Chemist showed that the sheep brains were actually cow brains.

The two audits published on Friday suggest that a mix up happened because pureed sheep and cow brains were stored next to one another in the same refrigerator.
The audit from Risk Solutions, a London-based consultancy, concluded that the most likely mix-up came in 1997, when technicians removed samples from the refrigerator for the mouse experiments.
"The most likely place, if a swap happened, was at the IAH," says Helen Wilkinson, author of the Risk Solutions study. She said that poor labelling of bottles might have been to blame: "The labelling didn't identify the species."

Old and decaying

The other audit, by the UK Accreditation Service, was less emphatic, but accused the institute of substandard labelling and record keeping.
Another possibility raised by the Risk Solutions report is that the samples got mixed up in 1992 or 1993, following the experiments for which the cow and sheep brains were originally collected.

Bostock is adamant that from the outset, he and his staff have had misgivings about the material because it was known to be old, decaying and possibly contaminated. He says that since there is now no way of finding out what happened, it's "time to move on to what's happening today".

Britain: Government suppresses report showing hospital patients face danger from Human BSE; 29 November 2001

The Labour government has suppressed a damning report into the procedures used by hospitals to prevent the spread of the incurable brain-wasting disorder variant Creutzfeldt-Jakob Disease (vCJD).
There are 111 confirmed or probable cases in the UK of vCJD, the human form of “mad cow disease” (Bovine Spongiform Encephalopathy or BSE). According to scientists, the total number of cases could be between several hundred and 150,000.
David Hurrell, an expert on the investigation team recently revealed to the BBC television programme *Panorama* that a top health official had ordered the destruction of all copies of the report. Hurrell says the report—now known in the health service as “the survey that didn’t take place”—showed that procedures to decontaminate surgical instruments were “barely adequate” in most of the hospitals investigated. He believes they are “an accrued result of 20 years or more of neglect”. Professor Michael Banner, chair of the Department of Health CJD Incidents Panel also told *Panorama* “it’s really quite absurd and unbelievable that the document has not been made available.”
In 1999, Health Minister Alan Milburn commissioned an urgent investigation into the methods used to decontaminate surgical instruments in 43 selected English hospitals. The move came after scientists warned that the biggest risk of vCJD being passed from person to person came from surgical instruments—particularly the re-use of surgical instruments used in brain, nerve and eye operations. Recent evidence suggests that lymph tissue—which produces the body’s white blood cells and is present in such organs as the tonsils and appendix—is also infective. There is additional concern in medical circles about the use of blood and blood products after experiments on sheep showed that blood can transmit the disease.
Milburn told parliament in November 2000 that he would publish the report in April 2001.
In a letter dated 26 September 2000, leaked to *Panorama*, National Health Service Estates Chief Executive Kate Priestley wrote, “In light of the somewhat negative outcome... there is a

need to ensure, at the express request of ministers, that the final version and earlier drafts remain strictly confidential... please undertake to destroy [all copies] and to check that no relevant files remain on your system."

Ministers refused to appear on the *Panorama* programme, telling the producers not to be "silly".

It should be remembered that at the same time as Hurrell's report was being destroyed, the government was drafting its *Response to the BSE Inquiry* [1] which boasted how the government was implementing a culture of "open government" and "the open sharing of information and research on all topics".

The irresponsibility of the government's suppression of the hospital decontamination information is further highlighted by the fact that the Scottish National Health Executive published a similar study in February this year. This shows there has been little research on decontamination procedures since an in-depth study was carried out more than 40 years ago. The Scottish study found that most of the hospitals were "deficient in a number of key areas... [giving] serious cause for concern". It also documents how decontamination often takes place in unsuitable environments, many instruments are in need of replacement or upgrading and there are few records of what instruments are used on patients. A significant minority of hospitals re-use instruments only intended for single use.

Scientists have known for decades that classical CJD can be transmitted to other patients after surgery on the nervous system. Many scientific studies say that the infective prion agent, believed to cause CJD, is remarkably resistant to conventional sterilisation and disinfection techniques. Recent research by Professor John Collinge at London University suggests instruments can become infected after five minutes contact with infected tissue. They remain infectious after normal decontamination procedures and can re-infect after 30 minutes contact.

In 1990, soon after the BSE epidemic in cattle began to escalate, records provided to the BSE Inquiry show government officials were discussing "appropriate decontamination procedures and blood products." It was another four years—"a quite unacceptable delay" according to the BSE Inquiry—before a guidance document on the risks of contamination to laboratory and hospital workers was published. It was not until April 1998 that the Advisory Committee on Dangerous Pathogens suggested that instruments must be destroyed—but only from those patients with symptoms of vCJD or suspected of having vCJD.

As researcher Andy Hill at the University of Melbourne points out, "The bottom line is that healthy cattle may harbour infectivity and never show signs of BSE. It is entirely possible that, in the same way, humans might be harbouring the disease at this sub-clinical level... we don't know how many people might be harbouring the disease." Brain surgeon Henry Marsh told the *Panorama* programme "The problem is the whole population of this country has been exposed to the BSE agent".

David Hurrell says his report "clearly indicated that it wasn't possible in many cases to rely upon decontamination as being an effective barrier to the transmission [of vCJD]." However, the government pressed on with its announcement of £200 million to improve decontamination procedures in January this year and the introduction of single-use instruments for tonsillectomy. Their introduction in other areas of surgery was rejected on the basis that the instruments were too "sophisticated" [i.e. expensive]. Subsequently, the government has sent a consultation document to clinicians asking them what instruments or blood products should be removed from use and when—12 years after the subject was first discussed according to official records.

On the *Panorama* website [2], Operating Department Practioner David Caldwell points out that operations on the throat close to the tonsils (laryngoscopy) are performed far more frequently than tonsil operations. They often result in damage to the tonsils yet the devices

employed are reused, often with the minimum of disinfection. He says the “consistency [of procedures] is illogical and the measures taken strike me as token gestures.”
In September this year, the *Sunday Times* newspaper reported that hospitals are having to warn thousands of patients about their operations. Research has shown that up to 41 hospitals unwittingly operated on patients incubating vCJD and then reused the instruments on other people. Medical staff have given transfusions to 22 people, and possibly thousands more, using blood donated by 15 people who later developed vCJD. Only blood from eight of the vCJD victims has been traced. Other newspapers report old surgical instruments being exported abroad.
It may be the case that the risk of spreading vCJD through hospital procedures is extremely low. However, by suppressing politically unfavourable information the government is sabotaging a scientific resolution of the problem and is denying the general public its right to be informed about a matter that raises critical public health issues. Or as David Hurrell put it, “At the moment what we have is essentially a dictated policy... with very little discussion going on among the professional groups that should be involved.”
* * *

Britain: Labour government accused of cover-up over BSE experiments; 26 October 2001

Farming and Environment Secretary Margaret Beckett has been accused of seeking to suppress how vital experiments concerning the safety of British lamb and mutton were botched-up. Scientists had hoped to determine whether deadly Mad Cow Disease (Bovine Spongiform Encephalopathy or BSE) has infected British sheep.
Most scientific opinion accepts that eating beef infected with BSE causes the fatal and incurable brain wasting disorder variant Creutzfeldt-Jakob Disease (vCJD) in humans. There are 107 confirmed or probable cases of vCJD in the UK. The total number of cases could be between several hundred and 150,000.
Four years ago scientists at the Institute of Animal Health (IAH) were commissioned by the Ministry of Agriculture, Fisheries and Food (MAFF) to check brains thought to have come from sheep that had died of scrapie in 1990-92at the height of the BSE epidemic. Scrapie is a disease similar to BSE that has been known for centuries but does not appear to affect human health. Since the early 1990s, scientists have become increasingly concerned that sheep diagnosed with scrapie may actually have BSE. By examining the brains, the IAH scientists hoped to find out if this was the case.
In December of last year, however, Professor Chris Bostock, the director of the Institute of Animal Health, became concerned that the sheep brains had been contaminated with cow tissue. The *Guardian* newspaper also reports that senior agriculture officials knew in early summer that there had been contamination, ruining the experiments. None of this was made public until early last month, when the government's BSE advisory committee hinted at a problem and suggested that all the brains used in the experiment should be retested using another method.
On September 28, the Department for the Environment, Farming and Rural Affairs (Defra), which replaced the largely discredited MAFF, published the government's substantive response to the report issued by the official BSE Inquiry in October 2000. Presenting this response, Defra Parliamentary Undersecretary Eric Morley claimed, "The culture of secrecy and protection of the food industry criticised in the [BSE Inquiry] report had disappeared." Media attention focused on the £120,000 compensation payments paid to the families of current vCJD sufferers. Little attention was paid to the two pages covering BSE in sheep and the government's *Contingency Plan for the emergence of naturally occurring BSE in Sheep in*

the United Kingdom National Flock. In section 2.12 of the *Response* are the words, "Currently BSE is not known to have occurred naturally in sheep. However, a scientific experiment is underway to test whether sheep infected with scrapie during the 1990s actually had BSE. Scientists are examining brain tissues collected from sheep killed at that time. The experiment is not yet complete, but preliminary results could be compatible with BSE having been in sheep at that time. However, scientific experts advising the Food Standards Agency have said it is not yet possible to draw conclusions from the research. The reasons for this arethe research is still incomplete; and there is a risk the sheep brain tissue being tested may have been contaminated with BSE-infected cow brains."

Elsewhere the government's *Response to the BSE Inquiry* says, "Ministers are fully involved in the decision-making process and are consulted on (or participate in) all important decisions. Ministers have also made clear that they expect decisions with public health implications to be referred to them in a timely manner."

Last week it was revealed that the brains had not just been *contaminated* by cow tissue. The scientists had, in fact, been using brains from *cows* and not sheep. This information prompted Beckett to issue a late night press release on Wednesday October 17, saying the tests had been ruined through "contamination" but avoiding any mention of the word "cow".

The following day, Professor Roy Anderson, head of Infectious Disease Epidemiology at London University who sits on the government's BSE advisory committee, revealed the full story. Scientists had been experimenting on cows' brains by mistake. The experiments, Anderson continued, have been "a great waste of time and effort and deeply misleading". He also blamed government for relying too much on the results from these tests and not carrying out other investigations and research. He suggested thousands more sheep should have been tested each year before they were sent for human consumption.

On Saturday October 20, Beckett was asked on BBC radio why the government had not tested the thousands of sheep that Anderson thought necessary. She replied, "Well, I'm in the same position as you are, and indeed Professor Anderson at this moment. I don't know the answer to that question either." She defended herself by saying, "I first found out about the experiment that had gone wrong on Wednesday afternoon."

By Monday October 22, Beckett was forced to make a statement to parliament, which included the incredible admission, "We have known *since the experiments began* that there were some doubts about whether the brains ... were cross-contaminated" [Emphasis added]. She also revealed, if even this can be believed given the penchant demonstrated by government bodies for using the wrong species in experiments, that only 180 sheep have ever been tested for BSE. The experiments, "have reached the point at which, if any of these scrapie cases was BSE, this might have become evident. It has not done so. However, it is too soon to draw firm conclusions from these ongoing experiments that can last several years."

If the experiments had shown that sheep had died from BSE and not scrapie, the consequences for human health and the farming industry could be far worse than the BSE crisis in cattle.

Recent experiments show that although BSE is largely confined to the brain and spinal cord in cattle, it spreads into far more organs and tissues in sheep, including meat/muscle. The current ban on the use of the brain and spinal cord from sheep and cattle would be insufficient protection. Officials from Defra say that if BSE had been found in these experiments then almost the entire 40 million sheep flock would have to be slaughtered. This would bring further devastation to an industry that has been decimated by BSE in cattle and more recently Foot and Mouth Disease.

The government has abandoned the Institute of Animal Health experiments. If the experiments were known to be flawed from the start, as Beckett now reveals, the fact the government did not pour millions into alternative research is criminal. There has been

virtually no testing of sheep and there is still no suitable test procedure to establish BSE in a live animal, years after the need for one was first identified. The government actually insisted on a special exemption from European Union rules requiring the compulsory testing of meat used for human consumption. The words "open government", "the open sharing of information and research on all topics", "joined-up government" and "the precautionary principle" that litter the *Response to the BSE Inquiry* are a mockery. Nearly 15 years after BSE was first identified in cattle, we are no nearer knowing if sheep are similarly affected or what the risk to human health is.

Britain: Big increase in human form of "Mad Cow Disease"; 11 September 2001

The incidence of variant Creutzfeldt Jacobs Disease (vCJD)—the human form of “Mad Cow Disease”—has increased 20 percent in the UK since last year. In his announcement last week, Professor James Ironside, head of the CJD Surveillance Unit in Edinburgh, said that instead of “a flat line, we are now seeing an upward trend that has been sustained for the past four quarters”. The total number of cases could vary between several hundred and 150,000, he added. Professor Ironside’s unit has released figures showing there are now 106 confirmed or probable cases of vCJD, the fatal and incurable brain wasting disorder in the UK. Most scientific opinion now accepts that the disease is probably related to eating beef infected with BSE (Bovine Spongiform Encephalopathy), or “Mad Cow Disease”.

Ironside also revealed that people in the north of Britain are twice as likely to get the disease as those living in the south. He thought this could be due to differences in genetic make-up, but was more likely to be the result of differences in diet.

Professor Tim Laing, from the Centre for Food Policy, told the BBC that differences in diet were a “class issue”. Cheaper meat products such as pies, sausages and burgers often contained the most infective tissues such as brain and spinal cord, before their use in the human food chain was banned. “Lower-quality meat products tend to be eaten by people on lower incomes, so in the north-south gap we might be seeing the beginnings of a class element to vCJD,” Professor Laing said.

Other scientists are sceptical that regional differences in diet are the cause of a north-south gap in the distribution of vCJD. But the resolution of this scientific question has been hampered by the actions of the food manufacturers, who for several years have refused to give statistics to epidemiologists about the use and distribution of mechanically recovered meat (MRM), a product widely used in meat pies, sausages and so-called “economy burgers” and which has also been included in soups and prepared meals. The MRM slurry is obtained by blasting cow carcasses with high-pressure water jets after removal of the prime cuts. Members of the government’s Spongiform Encephalopathy Advisory Committee (SEAC) complained last month that they had tried for five years to get the meat industry to provide the information.

The British Meat Manufacturers Association (BMMA) has said the information is difficult to obtain or is non-existent, although it seems only twelve premises processed MRM. A report in the *Independent* newspaper said the BMMA did carry out its own confidential survey in 1997, but the data was “lost during an office move”. The BMMA also says it raised the need for an official survey in 1997-98 after Labour came to power, but “it wasn’t done”.

The MRM information is vital for scientists such as epidemiologists trying to understand its role in the spread of vCJD. After a two-year investigation, the official BSE Inquiry, set up by Labour shortly after coming to office in 1997, said, it is “now clear is that this was the route by which infectious material was most likely to enter the human food chain.”

In 1989, the Conservative government banned the most highly infective tissue—brain and spinal cord—from human consumption. However, it still allowed the backbone to be used to obtain MRM, provided the slaughterhouses could guarantee 100 percent removal of the spinal cord. The BSE Inquiry pointed out that a Ministry of Agriculture report in 1990 on slaughterhouse practices "might have led one to expect such failures [of that guarantee]". Even if the spinal cord had been removed, other infective nervous tissue such as the dorsal root ganglia would be left behind.

The government was reluctant to ban the use of MRM, arguing that meat industry profits would be affected. At that time, over 5,000 tons of MRM were produced each year, worth about £3 million.

According to the BSE Inquiry, at its meeting in August 1994, SEAC agreed not to ban the use of backbones in MRM when Ministry of Agriculture officials gave assurances that the spinal cord was being removed. At its meeting in June 1995, a ban was considered again but was postponed; one reason given was that "the impact of prohibiting the use of spinal columns on the meat industry would be enormous". The BSE Inquiry also notes that despite assurances, some officials already realised there were "potentially serious failings" in the ability of slaughterhouses to completely remove the spinal column.

At its November 1995 meeting, SEAC learned that checks had found spinal cord contaminations on 17 separate occasions in 16 slaughterhouses, and recommended a ban on the use of cattle backbone in MRM, which the government finally implemented in December 1995. Backbones from sheep and goats were only banned in 1998. The production of MRM, mainly from chickens, still continues in enormous quantities.

The BSE Inquiry concluded that the eventual ban on using backbones to obtain MRM, "as far as preventing fragments of the spinal cord from getting into the human food chain was concerned, this was to a large extent a case of shutting the stable door" after the horse had bolted.

A related food safety issue also came to public attention last week. Doctor Richard Kimberlin, a member of SEAC for eight years, warned of the dangers of BSE in lamb. He accused the Labour government's new Food Standards Agency of playing a potentially dangerous "waiting game" by not implementing a ban on sheep offal. Kimberlin believes BSE may have passed to sheep in the 1980s, but it has been masked by scrapie, a Spongiform disease similar to BSE, but usually non-fatal to humans. Recent experiments on sheep brains suggest that some animals originally thought to have died from scrapie could actually have died as a result of BSE. Many scientists believe that BSE possibly originated from mutations that occurred in scrapie, when sheep tissues were used in the manufacture of cattle feed. The resulting BSE could then have passed back to sheep, and consequently to humans who ate mutton.

Professor Malcolm Ferguson-Smith, who sat on the BSE Inquiry, accused the government of ignoring its recommendations and feared the 16-volume report would become "a hugely expensive doorstop". He attacked the government's decision to hold the foot and mouth inquiries in private, saying the "same old gang" seems to be in charge of the new Department of Environment, Food and Rural Affairs, set up in June to replace the Ministry of Agriculture, and regarded by many critics as little more than a lobby for agribusiness.

Mad cow disease reaches Japan"; 10 September 2001

Japan has announced its first cow with BSE, the first native-born case reported outside Europe. It probably results from the importation of contaminated British feed exports in 1990 and 1991. It is also not likely to be the only mad cow in Japan.

In 2000, the European Commission is believed to have judged that Japanese cattle were likely to carry the disease, but Tokyo prevented the Commission from publishing its assessment.

However the Japanese agriculture ministry released a statement on Monday reporting that a single cow in the Chiba prefecture, east of Tokyo, has been confirmed with BSE. There has been no more official information, but the cow is said to be a five-year-old Holstein whose brain was tested after it showed suspicious symptoms.

Five is exactly the age you would expect if British feed imports caused infections that were subsequently recycled in Japanese cattle, says Marcus Doherr of the Swiss Reference Laboratory for animal diseases such as BSE at the University of Berne.

British export statistics show that Japan's first direct imports of British meat and bone meal (MBM) were in 1990 and 1991, when it bought 194 tonnes. Between 1992 and 1996 it bought a further 139 tonnes. It may also have bought British MBM from countries such as Indonesia, which imported thousands of tonnes.

Export boom

There was an export boom for British MBM after 1988, when Britain stopped feeding MBM to its own cattle because of the BSE risk. At that time the amount of infection in rendered British cattle also peaked. Britain did not start removing the most infected tissues, such as brain, from MBM sold outside Europe until 1992.
"**Japanese cattle infected by the British MBM in the early 1990s would have become maximally infectious themselves in the mid-1990s,**" says Doherr. The cow discovered in Japan would have been a calf then, and most likely to be exposed to infection in feed made from Japanese cattle. This could mean that the infection has been recycled widely in Japan.
That was the conclusion European Commission scientists reportedly reached in July last year, when they judged Japan's risk of BSE infection along with that of EU countries, the US and others.
"But Japan requested us not to proceed," says a Commission spokesperson. The Commission has now assessed some 25 countries, but only Japan has blocked disclosure of the result.

Recycled animals

Sources close to the Commission say the scientists concluded that Japan's British MBM and cattle imports, plus its rendering practices, gave it the same BSE risk as Germany. Japan stopped feeding European MBM to cattle last December, but still recycles its own animals.
Japanese agriculture officials did not test any cattle for BSE until this year, when they started random screening of a few hundred cattle. But they announced in August that they would start formal surveillance for BSE in 2002.
Observers suspect that the announcement of the first BSE case, coming so soon after the surveillance programme was announced, may be a deliberate attempt to cushion the blow to Japanese consumers. Japanese typically eat beef from animals over five years old - the most likely to be infectious.

vCJD vaccine a step closer to reality; 7 September 2001

Pioneering experiments in mice have brought vaccines against the human form of mad cow disease a step closer to reality. Through genetic engineering, **Swiss researchers made the mice immune to a similar disease called scrapie.**

News of the breakthrough coincides with warnings of sharp increases in cases of variant Creutzfeldt Jakob Disease (vCJD), the human form of mad cow disease. So far, 106 people have succumbed to the disease, but cases increased in number by 20 per cent last year. Epidemiologists have warned that thousands could eventually succumb, but no one really knows.

Given the uncertainty, a vaccine that neutralises infection would be invaluable.

"It does give hope," says Frank Heppner, a member of the Swiss team which developed the mouse scrapie vaccine at the University of Zurich's Institute of Neuropathology.

In the way

Heppner and his colleagues gave mice extra genes which enabled the animals to make antibodies against prions, the abnormal proteins which cause scrapie.
Prions are grossly misshapen forms of a normal protein called PrPC, which sits harmlessly on the surface of nerve and brain cells. But if prions come into physical contact with the normal protein, they turn it into a prion as well. Once tainted, cells produce vast amounts of prions, which kill brain tissue, spill out and spread the infection to neighbouring cells.
Heppner believes that the antibodies literally get in the way, masking the normal proteins from view and preventing physical contact. "Prions can't find their usual gateway to the cell," he says.
Genetically engineering humans to make antibodies is obviously out of the question, he admits. But he says that the breakthrough is an invaluable first step because it proves that vaccination can work. It might be possible to give protection simply by injecting antibodies.

Too late

Scientists had thought it would be impossible to vaccinate against vCJD. They feared that antibodies against the prions would mistakenly target normal prions on healthy cells as well, causing the body to attack itself. **The new work shows this does not happen.**
Heppner and his colleagues plan to inject antibodies into normal mice next, to see if this alone is enough to stem or prevent scrapie.
But even if the injections work, they might be too late for patients with obvious symptoms of vCJD. By the time of diagnosis, these patients' brains are already beyond repair. "Neurons can't be replaced, so it's a case of 'game over'," says Heppner.
However, a vaccine might be much more effective in patients who are infected but whose brains are still free of prions. Disease strikes only when the prions reach the brain, which can take many years. Several teams are working on a diagnostic test for vCJD in patients without symptoms, but as yet there is none available.

Special report the BSE crisis: 05 September 2001

As a scientist, it feels as if I have been holding my breath for 13 years while the BSE epidemic took hold, was exported to much of the world, and then the human form got going. Now at last, it looks as if breathing can start again: the good news is starting to come through. Only a few weeks ago things were still looking fairly dire. In the UK, we have had 181,294 cases of BSE. **It hit a peak in 1993 and we don't expect to see our last cases until after 2006.** The disease is now rising rapidly on mainland Europe with, for instance, seven cases in Germany in 2000 and 94 already this year. **We had exported 25,000 tons of the infectious**

meal and bone meal (MBM) - by then banned for feeding to cattle in the UK - to Europe in 1989, and consequently their BSE epidemics may not be over until after 2010.

In the UK, one of the major problems was that we did not know which cow was infected before it had any symptoms. As a result we ate six out of every seven of them. This represents over 800,000 infected cattle entering the human food chain and the population eating 50 meals each made of their tissue.

So far, 106 cases of variant CJD have been identified, with between 1,000 and 10m potential cases incubating the disease. We don't know who to treat, whose blood to avoid at blood transfusion, who represents a risk at surgery or dentistry, or which asymptomatic cattle to dispose of. A diagnostic test is urgently needed, but we would need to be able to look for less then 1,000 molecules of the infectious agent in blood or urine - less than one thousandth of the amount that can be sought by the best test available so far.

The BSE inquiry report last year showed how, through inadequacy rather than villainy, the government convinced itself that the risk to humans was minimal. Why, then, waste money on research?

Early in the epidemic it was difficult for scientific researchers to get hold of BSE tissue samples just to use for tests. The government refused to fund any research to look for treatment "because it would suggest to the population that there may be a risk to them".

In the early 1990s I was the only researcher looking for treatments for the disease and had to do the work in my garage. **One scientist, Iain McGill**, quit the government research group after warning of risks being taken. **He was told it would be illegal to tell anyone what he had found.** When vCJD appeared, the government still considered whether or not to tell the public.

But at last the hiding of information, blocking of research, denying of facts and damning of anyone speaking out in this field appears to be over.

A group in Switzerland has now found a way to increase the number of prion molecules in a sample - perhaps enough to be able to identify these triggers for vCJD. From **Israel we have data showing that urine contains lots of the prion molecules and that these appear long before any symptoms.** A London company can now look for 1,000 molecules of other proteins and is using this to look for prions. There is now an official supply of tissues and blood fractions to try out on any new tests.

Companies around the world have realised just how much money they could make if they had a test for vCJD in its incubation period - and ideas are appearing out of the woodwork. I would be surprised if we did not have one by this time next year. But what could doctors do then? Will it be like the test for HIV in the 1980s? "Thank you for donating your blood, Mr Smith, but I am afraid that you have vCJD and we will not be able to treat you."

In fact, a treatment is also on the way, too. In 1997 the UK government invited the pharmaceutical industry to a meeting with the BSE/vCJD scientists. Only three companies turned up - there was no money to be made, so why should they? In 1999 Laura Mannuelidis at Yale showed that an anti-leprosy drug, dapsone, increased the incubation period of scrapie

in mice dramatically. Last year, an anti-malarial drug, quinacrine, was demonstrated in the US to stop prions growing in cell culture.

The story of Rachel Forber, the British woman treated for symptomatic vCJD in the US with quinacrine, has just been published. A patient in London has also started the treatment. **There are now over 40 drugs that are active against prion infection** and we have some that are of adequately low toxicity to permit testing on patients.

We expect that some of these drugs and diagnostics will also be useful against Alzheimer's disease, a condition that is extremely difficult to experiment on with animals or in test tubes. So we may well find, in the end, that BSE had a silver lining. We might even end up with government that does not deny bad news and avoid research in case of what it may find.

Antibodies raise hopes of prion disease cure; 24 July 2001

Antibodies which "cured" mouse cells of scrapie have raised hopes that the human form of mad-cow disease will one day be treatable.

So far, 100 people have succumbed to variant Creutzfeldt Jakob Disease (vCJD). Thousands more could fall sick, warn epidemiologists, so a treatment is needed urgently.

But pioneers of the latest breakthrough warn against false hope. "**These are tissue culture cells, so we still have to find whether the same thing happens in animals, let alone in people,"** says Charles Weissmann, who led the research at the Medical Research Council's Prion Unit at St Mary's Hospital, London.

Weissmann grew mice cells in the lab and infected them with a strain of mouse scrapie to mimic infection with vCJD. **As expected, the cells began producing prions, malformed versions of a normal protein found on the cell surface, especially in brain cells.**

It is these prions, given the symbol PrP^{Sc}, which clog up the brains of people with vCJD, sheep with scrapie and cows with BSE.

But when Weissmann **exposed the infected cells to an antibody codenamed 6H4, it halted the production of the prion proteins. The cells remained "healthy"** for at least six weeks after the antibody treatment had been given.

Destroying the indestructible

As an unexpected bonus, prion protein that had accumulated beforehand disappeared as well. This was a surprise, as prion proteins are widely thought to be virtually indestructible.
"It means that, contrary to what people thought, PrP^{Sc} is not that stable," says Weissmann. "As it turns out, it is destructible ... and it's probably enzymes that degrade it."
The antibody works by blocking the normal version of the prion protein, called PrP^{C} and produced on the surface of cells. He thinks this lends weight to the theory that it is through physical contact with this normal version of the protein that the abnormal prion protein infects a cell and replicates itself inside.
To add further credence to this, Weissmann treated some of the infected cells with a bacterial enzyme that strips a cell of all its surface proteins, including PrP^{C}. As with the antibody, production of the prions dried up once PrP^{C} was no longer available.

Alzheimer's "vaccine"

Encouraging as the results are, **Weissmann warns that the treatment might not work in people. "The next step is to try it in mice," he says.**
Even if it worked in animals, researchers would need to develop different antibodies that mask the human PrP^c protein. Secondly, researchers would need to find a way of getting the antibodies into the brain, again, not a trivial task.
But he says that similar approaches to treat Alzheimer's disease have begun to yield interesting results. Elan Pharmaceuticals in Dublin, Ireland, announced on Monday progress with AN-1792, a "vaccine" against Alzheimer's disease.
The vaccine compound resembles amyloid protein, the substance in the plaques that affect the brains of people with Alzheimer's. The hope is that AN-1792 tricks the body's immune system into attacking and disposing of the plaque.
Elan claims that tests on 100 patients with mild to moderate Alzheimer's showed that some developed an "immunological response" to AN-1792. Elan is now planning tests in 375 patients in the US and Europe.

Journal reference: Proceedings of the National Academy of Sciences (vol 98, p 9295).

World Health Organisation says BSE is a major threat; 6 July 2001

Scientists last month warned that Bovine Spongiform Encephalopathy (BSE), or Mad Cow Disease, “has joined AIDS as a major health challenge facing the world.” A conference organised by the World Health Organisation (WHO) and the United Nations Food and Agriculture Organisation (FAO) concluded with a call for governments to “strongly consider” testing for BSE in cattle used for human consumption and imposing a worldwide ban on meat and bonemeal cattle feed (MBM).
BSE and its human equivalent, variant Creutzfeldt Jacobs Disease (vCJD) cause a fatal wasting away of the brain. The infectious agent is believed to be a mis-shaped prion protein, causing the development of lesions in the brain. There is no cure for vCJD, although Dr John Collinge, director of the new prion unit at Imperial College in London believes that “in the next five years we may be able to produce something that .... provides a treatment for this disease”.
The disease has already claimed 102, mainly young victims in Britain, three in France and one in Ireland. There are suspected cases in Hungary, Hong Kong and elsewhere. The charity, Wellcome Trust, estimates there will be about a quarter of a million cases of vCJD in Britain by the year 2040.
Samuel Jutzi, director of the animal production and health division of the FAO, warned the conference of the possibility that BSE could spread far outside the European Union. He said “It’s safe to say that eastern Europe may have imported sizeable amounts of risk given the sheer trade figures we have. Another area may indeed be the Near and Middle East.” A few days later the first country in eastern Europe—the Czech Republic—reported a case of BSE. Since the Czech government only banned the use of high-risk cattle by-products for human consumption in January 2000, it is clearly possible vCJD cases will start to appear there.
BSE was first recognised in Britain in the early 1980s. There have been 180,900 cases of BSE in Britain and officially there are still 1,500 cases a year. Changes in the rendering industry, and the increased use of MBM in cattle feed, are thought to have increased the spread of the disease.
In 1988, some time after MBM was suspected of spreading BSE, the Conservative government of Margaret Thatcher banned its use in feed for ruminants such as cattle, sheep and goats in Britain. However, the government allowed the export of MBM until 1996 when it

finally admitted a link between BSE and vCJD. Records show that over one million tonnes of MBM were exported from Britain to Asia between 1988 and 1996. According to Keith Meldrum, the government's chief veterinary scientist at the time, it was "up to importing countries to stop accepting our exports." For 11 years Britain exported the remains of BSE-infected cows to more than 80 countries where it was often repackaged and re-exported.

Where governments around the world have taken action against the BSE threat—and most claiming they are "BSE free" have not—the policies have been limited, uncoordinated and often unenforced. Protecting national business and agricultural interests by governments, industry and trade unions has been the major factor. The search for a suitable test and cure has therefore been delayed. The tests that are currently available are not sensitive enough to detect the small amounts of prions circulating in the blood (although two research groups are now developing tests) and it is still only possible to carry out tests on brain material where the prion is most concentrated. In Switzerland, where the first case of BSE occurred in 1990, *all cattle* are now tested for BSE after slaughter using a new rapid testing method on the brain. The older method appeared to show a dramatic decline in the disease in 1998, but with the introduction of the new method the following year there was a four-fold increase in cases of BSE recorded. This points to massive underreporting in every other country, and Britain in particular.

On July 1, new EU BSE Regulations came into force due to the spread of the disease to Ireland, Portugal, Switzerland, France, Germany, Spain, Netherlands, Belgium, Italy, Liechenstein and Luxemburg. Many of these countries had denied they had a BSE problem. Until tests proved otherwise, the German government declared the country to be BSE free. There have been 90 cases so far, 77 of them this year. In Spain, 48 cases has been detected—all but two of them this year.

The new regulations say that slaughterhouses must carry out a BSE test on all cattle over the age of 30 months that are used for human consumption and sick animals over 24 months. Some random tests of healthy cattle over 24 months must also be carried out. In Britain the government is only required to randomly test 50,000 cattle over the age of 30 months. The British Food Safety Agency (FSA) claim there is no need for any testing in Britain because of the national ban on the sale of beef from cattle over the age of 30 months. However, they will carry out the random testing in order to provide epidemiological evidence. The Agency said, "We support the right of other countries to test all cattle over 24 months, but in UK conditions we do not believe that testing under 30 months would provide additional public health protection." The FSA press release states that "in the UK, testing has until now been carried out primarily among animals aged over 30 months", but it does not say that the numbers tested were only 171 between January and May this year out of the European total of 3.5 million.

Professor Hugh Pennington, professor of bacteriology at the University of Aberdeen, condemned such complacency and omission. Pennington, who chaired the inquiry into the E.coli food poisoning outbreak in Lanarkshire, Scotland in 1996, said he feared "new food disasters on a similar scale to BSE... Nobody knows when the next food-borne bugs will arrive but they are evolving right now. I fear that without fundamental reform in the way policy makers get and use scientific advice there will be big trouble ahead." He continued that the Labour government is too secretive and criticised the link between science and industry.

Professor Richard Lacey, who was persecuted for his criticisms during the BSE crisis, says, "Blair continues to deceive. There are an unknown number of animals infected and people are still eating contaminated beef."

These criticisms of the Labour government are borne out by the foot and mouth epidemic that has resulted in the infection of 1,800 farms and the slaughter of 3.5 million animals in the UK. In 1997 the Labour government's Spongiform Encephalopathy Advisory Committee warned

against feeding pigs waste meat. Jack Cunningham, the Agriculture Minister said at the time, “processing certain types of waste containing porcine material and feeding it as swill to pigs will have to end.” But the use of catering waste as swill continued. The current epidemic of foot and mouth disease is believed to have started from this source.
More than 90 burial sites were used to dispose of all the cattle slaughtered due to foot and mouth disease. The burials included cattle over five years old, which should have been incinerated under the government’s own BSE regulations. The government has now ordered the carcasses to be dug up to protect water supplies from infection with the BSE agent—a rather belated attempt because most body fluids will have leaked out after two months.
Such practices underscore that the BSE Inquiry set up by Labour two years ago in response to public outcry was an entirely cosmetic exercise. Only last month European veterinary inspectors published a report of their investigation into the British meat industry. It said processed meat products “gives rise to serious concern” and checks on raw materials used for food were “weak or even non-existent”.

Global threat; 15 June 2001

The three biggest international agencies for health and agriculture want all countries to assess their risk of BSE, and to watch for the disease. They called on rich nations to help poor ones evaluate risks and "identify" precautions, but stopped short of calling for help with expensive BSE testing.

BSE is likely to have spread worldwide in the animal feed containing meat and bone meal (MBM) exported from affected countries, especially Britain. **New Scientist warned in February that disease could have spread globally.**

Thailand and Indonesia are most at threat, having imported large quantities of MBM, though Indonesia may have re-exported much of this to China. China denies any risk.

At a summit meeting in Paris, which ended on Thursday, the **World Health Organisation, the Food and Agriculture Organisation, and the World Animal Health Organisation declared that "ruminant MBM ... should not be fed in any case to ruminant animals".**

Sheep and goats

It is not always clear where imported feed really originated, and low levels of BSE in cattle can escape detection, so the agencies have urged all countries to double-check their risk. They called for better tests to detect undeclared ruminant protein in feed.
Countries at risk should also "strongly consider" testing livestock, the agencies recommended. This was how many EU countries discovered they were infected, but it is too expensive for most developing countries.
The agencies recommended precautions such as removing brain and spinal cord from sheep and goat meat, in countries where the livestock may have been given suspect feed.

> They said "research to date" shows pigs and poultry should be safe, without mentioning that research has been limited. But they called on scientists "to communicate new information about BSE and its risks as it becomes available, even though it may be unsettling to the public."

BSE / "Mad Cow Disease" spreading in Spain; 7 May 2001

Forty-three animals infected with BSE, or “Mad Cow Disease,” have so far been registered in Spain. According to official information provided by the department of agriculture and fisheries, 33 of these are concentrated in the north-west area of Galicia. The others are in Asturias/Basque Country (6 cases), Barcelona (2) and the Balearic Islands (2). The cases were reported between November 22, 2000 and April 3 this year.

Some experts believe that the number of BSE cases in Spain will rise to more than 250 by the end of this year. The most conservative assumptions predict that there will be a minimum of 3,500 cases in the course of this decade. However these predictions are based on the present level of known infections, which is about one positive case per thousand animals tested.

Scientists believe a prion protein agent, which produces holes or lesions in the brain, causes Mad Cow Disease. BSE and its human equivalent, variant Creutzfeldt Jacobs Disease (vCJD), lead to a “wasting away” of the brain, inevitably ending in death.

Human BSE has already claimed 92 victims in the UK, three in France and one in Ireland; most of these being mainly young people. No cases of vCJD have so far been notified in Spain. Although the mother of Javier Monge, who died last year, claims that her son showed symptoms of the disease. Monge had lived in the UK for 16 months and was admitted to the Doce de Octubre hospital in Madrid in May last year with symptoms that were compatible with CJD.

BSE was first recognised in Britain in the early 1980s (although reputable microbiologists consider it to have been present for two or three decades before that), and soon spread to Ireland. Changes in the rendering industry, and the increased use of animal protein in cattle feed are thought to have exacerbated the spread of the disease in the UK.

The European Union (EU) imposed a ban on British beef to supposedly prevent BSE from infecting cattle on the continent.

However, nearly twenty years on, the disease is now spreading throughout Europe. The first cases outside of Britain and Ireland were registered in Portugal and Switzerland in 1990, followed by France in 1991, Germany, Denmark and Luxembourg in 1992. At present, the other European countries most affected are Portugal (538 cases), Switzerland (366) and France (279), followed by Germany (55), Spain (43), Belgium (23), Holland (14) and Italy (7).

The Spanish government now stands accused of delaying tactics. The first Spanish cow reported with BSE symptoms was “Elvira,” which died in August last year. By October, the government knew it had been affected by BSE but did not confirm this until December. The family who owned the cow has accused the regional government in Galicia of having pressurised them for at least two months to keep quiet about the case.

But even more damning is the fact that for several years agriculture ministers in Madrid had blocked measures proposed by the EU to stiffen controls against BSE. Together with Germany, Denmark, Austria, Sweden, Finland and Greece (although many of these later changed their position), Spain refused to ban parts of the animal known to be the most infective—such as the brain, spinal cord, tonsils and intestines of animals over one year old—from entering the human food chain. This was the still case last June, when Prime Minister Aznar, in response to a question by the leader of the social democratic PSOE in parliament, demanded that people “should not generate alarm, since the situation in Spain was under control”. Ministers argued that to implement such a ban, which had been demanded by the European Commission since 1996, was a discriminatory measure against countries such as Spain. The ban already in place in the United Kingdom could not be extended to the rest of the EU, because countries like Spain were free of the epidemic, they contended.

Even the few controls that were introduced in Spain regarding the use of animal feeds containing meat and bone meal products were not enforced. Similarly, the ban on imported cattle from Portugal, where BSE had become more widespread, was being broken in hundreds

of ways. The border between Spain and Portugal is 1,215 kilometres long, it comprises an extensive territory marked often only by posts. Spanish abattoirs are used to slaughter many Portuguese cattle. Some Spanish farmers buy land in Portugal for their cattle to graze, because they enjoy better subsidies and other fiscal advantages there. Many have admitted they bought animal feed in Portugal, since it was much cheaper there.
Once the cases of BSE could no longer be hidden, the Spanish government said it would finance 40 percent of the emergency measures, seek EU funding to match its contribution, and ask the regional administrations to foot 15 percent of the bill, with farmers contributing the remaining 5 percent. However, it has levied a special tax on all meat products—not just beef—to pay the estimated 60 billion pesetas ($321 million) cost of dealing with the outbreak of BSE.
Spain is presently poorly equipped to face a BSE epidemic. Few veterinarian laboratories are equipped to carry out tests for the disease, and there are only five incinerators capable of disposing of contagious material, and one of these is out of commission. Breeders complain of too little compensation and there are fears that falsified ear tags are being used to conceal the true age and provenance of cattle. In Galicia, cattle rustlers have continued to trade across the border with Portugal. The regional authorities are struggling to comply with new EU directives on the testing of cattle and for the disposal of hazardous animal waste. BSE infected carcasses can often be found in the countryside, or buried in shallow graves. The majority of stock breeding companies are not complying with the regulations to control the disease. Of 2,500 inspections carried out up to January 15 this year, 2,000 infringements of the regulations to combat BSE were reported and at least 7 people were imprisoned as a result.

Moreover, Spain is divided into 17 autonomous regions, making the co-ordination of the anti-BSE measures and controlling the epidemic extremely difficult. The crisis has already claimed several political scalps. Castor Gago, Agriculture Secretary in Galicia, resigned after it emerged that the regional government was throwing diseased carcasses down a mineshaft, instead of disposing of them properly. Farmers have blocked slaughterhouses and meat packing plants across Spain to protest against the lack of government help to deal with BSE.

Prime suspect; 17 April 2001

An African antelope imported into a British safari park in the 1970s was probably the origin of mad cow disease, says a New Zealand epidemiologist.

Roger Morris of Massey University has used computer modelling to analyse dozens of possible explanations for BSE. "**The one that comes out top of the list is the African antelope hypothesis," he says.**

African antelopes are susceptible to spongiform encephalopathy, says Morris. He thinks a single infected animal used as feed for cattle could have triggered BSE. The BSE epidemic led to the slaughter of millions of cattle and the human form of the disease, vCJD, has killed more than 90 people so far.

"**The area of Britain where BSE started is the area where safari parks started in the 1970s," Morris told AFP.** "I've got evidence that every step in the sequence could have occurred."

Spontaneous mutation

But other experts are not convinced. The main alternative suggestions blame scrapie-infected sheep being fed to cattle or a spontaneous genetic mutation in cattle.
The antelope theory "is not the most likely hypothesis, given what happened with the BSE epidemic," says John Wilesmith, head of epidemiology at the UK's Veterinary Laboratory Agency, who has worked with Morris. But he adds: "**There are lots of complex theories for BSE and they're all worthy of examination."**
Nora Hunter of the Neuropathogenesis Unit at the Institute for Animal Health in Edinburgh agrees the idea is interesting but says: "There are numerous other possibilities and it will be very hard to prove."

Single strain

A wildlife source of BSE-causing prions would explain why the epidemic in the UK was unique and caused by a single strain of prion, Morris says.
The evidence "strongly favours" the idea that a wild animal of some kind was responsible, he says, and while an infected antelope is the most likely cause other animals, for example lions, can also develop a TSE (transmissible spongiform encephalopathy).
One difficulty with Morris's theory is that a wild antelope suffering from a TSE has never been identified. But this is not surprising, he says, as an afflicted antelope would rapidly fall prey to lions or hyenas.

Counting sheep

Morris **discounts the mutation hypotheses, which was favoured by the UK's official inquiry into BSE, published last year**. If a spontaneous mutation was to blame, "you would expect BSE to have cropped up sporadically in the past in countries with much larger cattle populations," he told **New Scientist** in November 2000.
The inquiry report rejected suggestions that changes to the way sheep are rendered allowed scrapie to cross to cattle and Morris agrees. Other countries have scrapie and similar rendering practices to Britain but none has had a native case of BSE, he points out.
However, **Wilesmith favours** the hypothesis. "**The factor that distinguishes Britain from lots of countries is that we have a large sheep population. Our ratio of sheep to cattle is uniquely high. A scrapie-like agent seems the most likely explanation,**" he says.

Pressure cooker

One legacy of the BSE epidemic is the difficult task of destroying the infectious prions from BSE-infected carcasses, but a Scottish company believes they have now found a way.
Prions resist standard sterilisation but Waste Reduction Europe has adapted a US technique, which involves pressure-cooking carcasses with sodium hydroxide to 1,500°C.

This destroys the prions and converts the carcasses into an aqueous solution that can be safely disposed of in a sewer, the company says.

More mad cows; 5 April 2001

The "highly likely" presence of BSE in eastern Europe, announced this week by the European Commission, means nearly a fifth of the European Union's meat imports have been coming from suspect countries that have applied no BSE safeguards, **New Scientist** has learned.

These countries may now only sell the EU meat that has been subjected to some - but not all - of the safeguards the EU requires of its own meat producers.

The Commission's risk assessments were based on whether the country imported cattle or meat from countries with BSE, and on whether cattle were recycled as feed, the route of BSE transmission (New Scientist, 10 June 2000 p 4).

The Commission used this method last year to conclude that countries such as Germany, Spain and Italy were "highly likely" to be infected - all have now declared BSE.

Ten countries judged "highly unlikely" to have BSE will be excused from these precautions. The Commission wants Brazil excused as well, even though it imported more than 6000 potentially infected cattle from Europe and fed cattle remains to cattle until February 2001. Last year Brazil exported 32 per cent more meat to the EU than the total from all the eastern European countries assessed.

Spinal cord

The countries judged to have BSE must now ensure there are no high-risk tissues, especially brain and spinal cord, in meat they sell the EU. The Commission says its verdicts also "should be of considerable interest to the public health authorities" wishing to protect their own consumers.
Meat from infected countries will not be subject to other EU safeguards, such as the ban on feeding cattle to cattle, or tests for BSE in abattoirs. Those tests became mandatory in January in the EU and by March had kept 38 mad cows off supermarket shelves.
The Commission thinks infection is "highly likely" in six candidates for EU membership - Poland, Hungary, Slovakia, Cyprus, Estonia and the Czech Republic. It "cannot be excluded" in India, Pakistan, Mauritius and Colombia. The countries deemed safe include Argentina, Australia, New Zealand and Botswana. The Commission will assess 17 more countries in May, including China, Thailand and Israel.

The five eastern European candidate countries exported 164,000 tonnes of meat to the EU last year, according to Eurostat, the EU statistics service in Luxembourg. That is small compared to a domestic production of 35 million tonnes. But in the EU, infected countries discard high-risk tissues.

Slow down; 2 April 2001

Two separate laboratories have discovered that a particular immune process helps prions cause disease. **Interrupting this process, in one case with cobra venom, delayed or even blocked prion disease in mice.** The observation might lead to a treatment for CJD - none exist at present.

Prion infections generally start when the victim eats or is injected with material from an infected animal. Eventually the deformed prion accumulates lethally in the brain. To get there, prions must first replicate. In scrapie, the best-studied prion disease, this happens in follicular dendritic cells (FDCs) in the spleen.

FDCs normally collect foreign molecules tagged for destruction by the immune system. One tag consists of blood proteins called complement. If a complement tag is needed for prions to invade FDCs, blocking complement might block that too.

Adriano Aguzzi and colleagues at the University of Zurich, and Neil Mabbott and colleagues at the Neuropathogenesis Unit of the Institute for Animal Health in Edinburgh, interrupted the immune systems of mice in various ways, then injected prions derived from scrapie.

Mice genetically engineered not to produce various components of the complement system, especially a circulating protein called C1q, or the FDC receptors for C1q, either did not develop scrapie or developed it later than mice with intact complement.

The Edinburgh team also suppressed the complement system temporarily with a toxin from cobra venom. Five days of suppression at the same time prions were injected was enough to delay the onset of disease by nearly a month, which is significant in mice.

The blockage could be overridden with larger doses of prions, suggesting that the prion can use other paths to invade the brain. But both teams are optimistic that the importance of complement in the early stages of infection suggests potential therapy.

Maurizio Pocchiari, of the Istituto Superiore della Sanitá in Rome, cautions that the importance of various invasion paths could differ with different prions and hosts. Also, complement may not be so important in humans and it is involved immediately after infection. This means any therapy would have to be early, difficult when incubation times are long.

But it may already be known to be effective. **The main compounds now being studied as treatments are glycosaminoglycans, and similar polyanionic compounds, which delay or block scrapie for reasons unknown.** One of the things they do, the Edinburgh group notes, is block the complement protein C1q.

More at: **Nature Medicine** (vol 7, p 485 and 488)

Like lambs to the slaughter; 31 March 2001

What if you can catch old-fashioned CJD by eating meat from a sheep infected with scrapie?

Four years ago, Terry Singeltary watched his mother die horribly from a degenerative brain disease. Doctors told him it was Alzheimer's, but Singeltary was suspicious. The diagnosis didn't fit her violent symptoms, and he demanded an autopsy. It showed she had died of sporadic Creutzfeldt-Jakob disease.

Most doctors believe that sCJD is caused by a prion protein deforming by chance into a killer. But Singeltary thinks otherwise. **He is one of a number of campaigners who say that some sCJD, like the variant CJD related to BSE, is caused by eating meat from infected animals. Their suspicions have focused on sheep carrying scrapie, a BSE-like disease that is widespread in flocks across Europe and North America.**

Now scientists in France have stumbled across new evidence that adds weight to the campaigners' fears. To their complete surprise, the researchers found that one strain of scrapie causes the same brain damage in ...

Britain: Report links CJD cluster to local farming and butchery practices; 27 March 2001

An official investigation of a cluster of five deaths from variant Creutzfeldt Jacobs Disease (vCJD) in the village of Queniborough concludes that local farming and butchery practices were the most likely source of the infection.

In the UK there have been 95 confirmed or probable cases of vCJD, the fatal brain wasting disorder related to BSE (Bovine Spongiform Encephalopathy) or Mad Cow Disease.

Leicestershire Health Authority carried out the investigation into the vCJD cluster in Queniborough, a village about 100 miles north of London with a population of only 2,297. Three of the victims—Stacey Robinson (19), Glen Day (35) and Pamela Bayless (24)-all died within months of each other in 1998. In May 2000, a 19-year-old man who lived nearby died, followed by 24-year-old Christopher Reeve last September.

Pamela Bayless's father Arthur said when he noticed how “Glen, Stacey and Pam all died within months of each other, I spoke to Glen's dad and we discussed how strange it was that it was all in Queniborough. It's such a rare disease.” He campaigned for an inquest into his daughter's death, but without success. Then in October 1998, the local newspaper, the *Leicester Mercury*, reported that three people had died in Leicestershire, two in the same village, and contacted the Health Authority for comment. The official response in 1999 said the deaths were a coincidence and posed “no cause for alarm”. The investigation only came about due to the insistence of the families of the young victims.

The Health Authority's report concludes, “The people who had vCJD were exposed to the BSE agents through the consumption of beef which had been processed from butchers. There was a risk of cross-contamination of bovine brain material during the boning and cutting process in those premises where the skull was split to remove the brain”. It suggests a moderately high incidence of BSE in the area originated in the mid-1970s as a result of farmers giving their cattle meat and bonemeal feeds containing recycled animal tissues. In addition, the cattle incubated the disease longer because they ate bonemeal from 6 days old, rather than the more usual 6 months, and were slaughtered later because they were slower-growing Friesians.

It was also usual practice in the early 1980s before BSE was discovered for small abattoirs in the area to sell cattle heads to local butchers who would split the skulls, cut out the brain—the most infectious organ containing BSE-and remove the remaining meat using the same knives they employed to do their other butchery. Thus local farming and butchering practices created a higher than usual risk of meat being contaminated with the BSE agent, the report concludes.

There have been conflicting reactions from several leading scientists to the Leicestershire report.

Professor Roy Anderson, an expert in BSE epidemiology, says the investigation has come to "a very plausible explanation," and was important for establishing an incubation period for the disease in humans of 10-16 years. However, "It is important not to over-interpret this cluster. It is significant, but it is only five cases," Anderson said.

Professor John Collinge, a member of the committee advising the government on BSE warned: "For me, the main finding from this report is that the significant exposure appears to pre-date 1985. That sent a little chill down my spine, certainly. It fits with our estimates that we have been making of the likely incubation periods of BSE in humans.

"The cases we are seeing at the moment are by definition those with the shortest incubation periods and the average incubation period could well be in the region of 30 years, Collinge told the press. "The upper limits of the modelling at the moment are in the region of one to two hundred thousand—that is one extreme of the possibilities—but we may see thousands, or tens of thousands."

Professor Hugh Pennington, professor of microbiology at Aberdeen University investigated the E.coli bacteria deaths resulting from contaminated meat in Scotland two years ago. Pennington said the report provided "a very plausible story and underlines what we know already, but does not explain why there was a cluster, because I do not think what they were doing was unique. It also does not explain why the victims were so young. It is very important data and very useful to have, but it has not unlocked the secret of CJD."

The most critical response came from Professor Richard Lacey, who was vilified by the then Conservative government and the media when he first exposed the BSE crisis and its implications for human health. Lacey said the report has scapegoated local butchers without addressing the real causes. He told the BBC, “They have no idea, it is just guess work, speculation. The aim is to reassure, rather than get at the truth. This has been the whole basis of CJD over 15 years—not to get at the truth, but to reassure in the short term.” He pointed out that vCJD is difficult to catch through the oral route (i.e. by eating contaminated meat), "It is not clear exactly how it spreads, it could be more than one way."

This dissent and caution among experts in the field of BSE is in sharp contrast to the official “spin” that has been put on the Leicestershire report, which came across more as a public relations exercise designed to reassure the villagers of Queniborough. The presentation of the report gave an overall impression that the deaths of five young people in the village were due to a series of unlucky coincidences.

But the farming and butchery practices in Leicestershire were not unique. Farmer's wife Margaret Winterton who has lived in the village for 18 years said, “I don't think it is the answer. I think a lot more investigation is necessary. I think the butchering practices that they have explained have been carried out all over the country—so why is there the cluster here?”

In response to the report's finding that butchery methods had played a role, local butcher David Clarke said, “You are talking about something from the Eighties, not something related to the present time.”

But ever since the emergence of BSE in cattle, the main thrust of government policy has been to protect the profit interests of the meat industry, something that has not essentially changed even when a direct link with vCJD in humans was admitted.

It is reported that health officials are also investigating the death of three people with vCJD in the Yorkshire village of Armthorpe, and of two men who lived within 250 metres of each other in Greater Manchester.

Mad cow USA; America denies having BSE, but has yet to prove it; 10 February 2001

America denies having BSE, but has yet to prove it

AS BSE casts its menacing shadow across much of the globe, the US is becoming increasingly nervous that its $400 billion beef industry may not escape. Officials have taken precautions to keep BSE out. But New Scientist has established that even if the US has as high an incidence of BSE as France, where it has sparked a health and farming crisis, American surveillance efforts would not spot it.

The US imported a mere 44 tonnes of British meat and bone meal (MBM) before 1996, plus 126 cattle that escaped a subsequent round-up and could have ended up as feed. But last year, the European Commission's scientific advisers warned that any infection in those imports would have been spread and amplified by American rendering and feeding practices (New Scientist, 10 June 2000, p 4).

American officials strongly deny this. "**We have no BSE," says Linda Detwiler**, who chairs the ...

Tomorrow the world; Have contaminated feed experts spread BSE across the globe?; 10 February 2001

Have contaminated feed exports spread BSE across the globe?

BSE may be about to go global. **Official British figures show that over 80 countries imported animal feed from Britain that was probably infected with mad cow disease** (see Map). And according to the UN, the rest of the European Union also trebled exports of potentially contaminated feed during the 1990s to non-EU countries.

Some of the biggest importers, such as Indonesia, Thailand and Russia, may now have the infection. The World Health Organization is holding an emergency meeting on the problem next month.

South-East Asia appears to be at most risk from infected meat and bone meal (MBM) exported by Britain between 1980 and 1996. Indonesia imported 600,000 tonnes during that time, Thailand imported 185,000 tonnes and Taiwan and the Philippines imported over 45,000 and 20,000 tonnes respectively.

Meat production in the region has boomed over the past decade (New Scientist, 18 March 2000, p 32).

Germany utilises BSE crisis to implement EU plans to restructure agriculture; 31 January 2001

With no let-up in the news of new BSE cases in Germany, the government is using the present indignation and disconcert in the population in order to implement a radical change in agricultural policy. The routine invocation of “consumer interests” is only the welcome cover for this project.

When Chancellor Gerhard Schröder (Social Democratic Party—SPD) boasts about his own common sense agricultural policy being rooted in the soil, and which should be thought of proceeding “from the shop counter”; and when he announces the end of large-scale farms in favour of small-scale agricultural production—to protect the consumer—then caution is required. When in the last two years has the SPD-Green coalition government ever placed the interests of industry, i.e., of large-scale enterprises, below those of the broad mass of the population?

In agricultural policy as in all other questions, an enormous gulf exists between the German government's words and deeds. Firstly, a more careful investigation of the facts disproves the government's campaign to lay blame exclusively at the feet of “harmful” large-scale farms (Schröder: “the promotion of family-run farms is in any case correct.”). So far in Germany, it is above all such smaller family-run farms that have suffered from BSE, and only recently has it beset one of the large-scale agricultural enterprises.

“BSE also occurs in ecologically-bred cattle, because the infective agent is not interested in the agricultural philosophy of the farmer,” says Udo Pollmer, scientific director of the European Institute for Food and Food Sciences in an interview with *Der Spiegel* magazine. Whether it is exemplary farms in Lower Saxony, which have never used animal-based feeds or even Swiss “eco-farms”, which supplied their cattle with self-cultivated plant fodder, the farms that have so far been affected are precisely the ones that the new agricultural policy is apparently aimed at creating.

Besides, a closer look at the changes announced in agriculture shows that, contrary to government communiqués, they will continue to strengthen large agribusiness. In March 1999 the European Union (EU) agreed *Agenda 2000*, which is now being presented as a decision to reverse agricultural policy along ecological lines, and which therefore has “finally” to be implemented. *Agenda 2000* envisages, among other things, the uncoupling of all production-based subsidies (i.e., those purely based on quantity) making them dependent instead upon environmental and quality standards. That is also the threadbare justification used in the present campaign in favour of “eco-farming”.

According to the criteria of “species-appropriate” and “land-related” animal husbandry, aimed at both in *Agenda 2000* and advocated in a recent seven-point program from the Health and Agriculture ministries, cattle may only be given plant-based feeds, which must largely come from their own farm. This would require pastureland of one hectare per cattle; a ratio that cannot be found on any average family farm, let alone the large-scale enterprises that engage in mass animal husbandry. In contrast to such large-scale operations, the land purchases required for farms that want to continue receiving subsidies cannot be afforded by small family concerns. Also the changes to the stalls in keeping with species-appropriate methods can cost even a small to medium sized operation up to $189,000. The effect of these two modifications alone would mean further “farm deaths” and strengthen the industrialisation of agriculture.

The number of farms in Germany has already dropped from over 1 million in 1970 to 429,000 today. At the same time, the average farm size of 11.7 hectares in 1970 has risen to 29 hectares in western Germany, and 201 hectares in the old East Germany (where large-scale cooperative farms were the rule). According to the government's *Agrarian Report 2000*, between 1998 and 1999 the number of farms sank by 5 percent. In the last 30 years the number of people employed in agriculture has halved from 2.7 to 1.43 million in 1999.

Despite enormous subsidies provided by the EU, the incomes of small farms sank continuously (for example, the average profit of a farm in North Rhine Westphalia sank from $576 per hectare in 1995/96 to $377 per hectare in 1998/99). This will hardly change given the sharp reductions in subsidies foreseen by *Agenda 2000*.

In drawing up *Agenda 2000* about one year ago, the EU governments had certainly not decided upon a program to combat the BSE crisis, which was handled at that time as a “purely British” problem, nor had they decided to turn to eco-farming based on small family farms. Rather the conflict-prone negotiations surrounding *Agenda 2000* were firstly concerned with preparing the European agrarian sector for the extension of the EU to the East. The addition of large-scale agrarian producers like Hungary, Poland and the Czech republic meant previous subsidy practices would have exploded. For Germany alone, carrying on with the old regulations would have meant an additional burden of over $6.6 billion annually.

Secondly, it also concerned improving the room for manoeuvre in negotiations with the World Trade Organization (WTO). The EU had already promised the WTO it would end its present subsidy practices by 2003 at the latest. The realisation of *Agenda 2000* would enable the EU to adopt “an offensive negotiation strategy”, as it was described a year ago. The EU's subsidy practices had been attacked by other food and animal feed manufacturers, above all in

the United States, because the European Union, by giving its farmers highly subsidised goods, provided them with an “illegal” advantage against their competitors on the world market.
The aim was to find a way out of Europe's confused and historically derived agricultural situation. Significantly, the reconstruction of European agriculture after the Second World War had lasted longer than that of industry, with the result that the economically backward agricultural methods of small family farms could gain a foothold again. In 1960, some 20 percent of all employed persons in the European Economic Community (EEC—the forerunner of the EU) worked in agriculture. At the same time, however, more intensive industrial production methods were being introduced into European agriculture. In addition, world prices for agricultural products sank, particularly due to an enormous rise in agrarian exports from the US. Most European governments reacted to this development—production above national requirements, falling world prices and a fifth of all persons employed in agriculture—with price and import controls, as well as by introducing subsidies for farmers. This led to the awful pictures of the meat and butter “mountains” and the wine and milk “lakes”, which had to be destroyed because they were too “expensive” for the world market, while hunger daily claimed thousands world-wide.
Under these conditions of protectionism, Europe rose to become the world's largest agrarian exporter, leading to sharp conflicts within the EU and with the international competition, in particular with the US. From the mid 1980s, it was this intra-European and international competition that led the agrarian industry to increasingly rely on so-called mixed fodder (blends of vegetable and animal meals), and so unleash the BSE epidemic among cattle and humans. In Germany alone, the production of meat and bone meal in 1999 amounted to 670,000 tons. The same year, the production of mixed feeds from 526 manufacturers (in Germany alone) rose to 19 million tons.
Like all other branches of industry, agriculture is a global system, and accordingly faces the same pressures from the world markets to diminish all restrictions and subsidies. However, agricultural subsidies form the basis of existence for the majority of European farmers. Their reduction will cause a social catastrophe, in particular in the poorer EU countries.
The recently resigned German Secretary of Agriculture, Karl-Heinz Funke (SPD), himself a farmer, shrank from introducing *Agenda 2000* too quickly, not least due to the effects on farmers. This is now to be pushed through by Renate Kuenast, the Green minister who has replaced him, who is not known to have any links with agriculture. Moreover, the Greens still enjoy a reputation as an “ecological party,” and this will be used to present the changes in agriculture as being in the interest of the small “eco-farmers” and the consumer.
The eco-farmers may now be rejoicing at the prospect of the millions of euros to be spent implementing *Agenda 2000*, but their faith in such government propaganda will not protect them from being displaced by agribusiness, just as it is happening to conventional small farms at present.

Consumer protection by ecologically-based agriculture and animal husbandry is not synonymous with economically backward small-scale farming as in pre-war times. It is not the technological and scientific developments in agriculture that are responsible for the recurrent food and environmental scandals, which cost innumerable human lives, but their use exclusively to maximise profits.

BSE/"mad cow disease" crisis spreads throughout Europe; 23 January 2001

Cases of BSE have now been identified in 10 of the 15 European Union (EU) countries, as well as Switzerland and Liechtenstein, which are not members. Although incidences are still

relatively few in number, the discovery of the disease across the continent has had a dramatic effect on beef consumption, which has fallen by 27 percent across the EU.

Along with a rise in the number of cattle infected with BSE (Bovine Spongiform Encephalopathy), the number of people who have died from its human equivalent, variant Creutzfeldt Jacobs Disease (vCJD), is also growing. By the end of last year nearly 90 people had either died or were dying from the fatal brain-wasting disease in the UK; with the yearly number rising from 15 in 1999 to 25 in 2000. A further six suspicious cases are also under investigation. Deaths from vCJD have also been reported in France and Italy.

France

Over 160 cases of BSE were diagnosed last year, more than five times as many as the 31 cases the previous year. The government has also admitted there are some 50,000 “mysterious deaths” in cattle every year. As a result, beef sales have dropped by more than 25 percent and beef has been removed from the menu in school canteens in several French cities.

Besides the health concerns, farmers have also protested as the shortfall in BSE testing facilities has led to large stockpiles of beef building up that cannot be sold until it is passed fit.

The crisis could yet have far greater political ramifications. Police raided three government ministries last week on the orders of an investigating magistrate. Judge Bertella-Geffroy ordered the raids to seek evidence that senior officials knowingly allowed consumers to risk exposure to vCJD. She launched her investigation following the lodging of official complaints of “poisoning” by the families of two French vCJD victims. Judge Bertella-Geffroy had also led the inquiry into the HIV-tainted blood scandal in the 1980s, when around 4,000 to 5,000 people, many of them haemophiliacs, were infected with supplies from the national blood bank. Former Socialist Party Prime Minister Laurent Fabius and two other ministers faced manslaughter charges for their role in the scandal. (A specially created court later acquitted them.)

Sales of MacDonalds burgers in France have been hit by the growing BSE crisis, down by 47 percent over the same week in November 1999. With profits for 2000 down, the global burger chain has embarked on an extensive advertising campaign across the continent, where it makes fully 25 percent of all its sales, to try and maintain its market position.

Ireland

The number of cases of BSE reported in Ireland is rising. There have been 580 confirmed cases since 1989, with 149 last year alone.

A number of abattoirs have been discovered leaving high-risk material on carcasses passed for human consumption. The spinal cord, brain, and other offal are potentially much more infective than other parts of the animal—by a factor of up to 1,000. An EU ban on “specified risk” materials means they have to be removed before the meat can enter the human food chain.

However, an audit of 96 of the country's 360 abattoirs found 18 (nearly 20 percent) that were not complying with the EU regulation, and as a result three were closed down.

BSE-infected carcasses and the high-risk material removed in the abattoirs are supposed to be destroyed in special high-temperature incinerators. However, Irish farmer's estimate that three more incinerator plants would be required to process the quantities of material presently accumulating and to prevent a dangerous build up.

In Northern Ireland, 41 tonnes of German beef was seized from two Newry processing facilities when it was discovered to contain spinal cord, banned since last October. The meat was apparently being processed in Newry prior to shipping back to Germany.

Germany

The first case of BSE in a domestically bred cow was reported in November last year. Since then a further 21 cases have been confirmed.

So far, the political fall-out from the BSE crisis has been greatest in Germany. Earlier this month, Health Minister Andrea Fischer (Green Party) and Agriculture Minister Karl-Heinz Funke (Social Democratic Party) resigned, as the number of BSE cases rose. Funke had admitted failing to take any action, despite being presented with a report in March 2000 predicting BSE in the German herd.

According to *Die Welt* newspaper, in a letter to the EU Consumer Protection Commissioner, Germany's new Agriculture Minister Renate Künast has admitted that a German BSE epidemic on the scale of the UK's could be possible.

The new Consumer Protection Ministry (specially created as a result of the crisis) has confirmed that the "quick" BSE tests introduced in Germany for all cattle over 30 months old destined for human consumption do not always provide conclusive results. Newsweekly *Der Spiegel* said two of the new tests had returned negative results, although the cow was infected with BSE. The laboratory that conducted the tests has blamed the poor quality of the samples it was provided.

A poll last week showed that more than 50 percent of those questioned had little or no confidence in the safety of German beef products. The polling organisation GfK said beef consumption had plummeted, with households buying 59 percent less beef in November 2000 than in the same period the year before. In the northernmost state of Schleswig Holstein, where the first BSE case was discovered, the figure was 80 percent.

As a result, some 5,000 workers in the meat processing industry have been placed on short-time working. Major meat processors Nordfleisch and Moksel report sales down 20-30 percent in November last year, leading to cuts in working hours in their plants. The crisis has badly hit German *wurst* or sausage sales. Christian Zorn, one of the country's 15,000 medium-sized producers said that if sales fell any further, some 10,000 butchers could be out of a job.

Italy

Last week, Italian Prime Minister Giuliano Amato appealed for "calm" as it was expected tests would confirm the country's first home-grown BSE case. Despite his efforts to shore up the Italian beef industry, domestic meat sales have dropped by 40 percent. As elsewhere, the initial reaction from the beef industry and government has been to claim it was a "foreign" problem. Cremonini, Italy's largest meat processor, boasted "Italian meat—safe and guaranteed" only days before a case of BSE was confirmed at one of its slaughterhouses this month.

Health Minister Umberto Veronesi tried to play down the implications of the BSE crisis, saying "Italian meat is safer than it was five or 10 years ago and that is a certainty for consumers".

Spain

The first BSE case was reported in November last year. The government, which has now established a special committee to monitor BSE, has since admitted the disease may be more widespread than it first admitted. Five confirmed cases so far have also caused beef sales to drop by 25 percent.

Farmers held protests last week in 11 of the country's autonomous regions, calling for compensation. Regional authorities are struggling to cope, with a shortage of veterinarians trained to identify the disease and only two national laboratories able to conduct the new BSE tests ordered by the EU. There is also a severe shortage of incinerating plants to destroy potentially contagious carcasses and other material. The only such plant in Galicia, one of the country's largest meat producing regions, broke down last week. Agriculture Secretary Castor Gago then ordered cattle carcasses awaiting incineration to be thrown down a disused mine shaft. This practice was only stopped when local residents complained of the stench. In Avila, 100km northwest of the capital Madrid, two suspect carcasses were found on a local rubbish tip.

The normally pro-government *El Mundo* newspaper has called for the Health and Agriculture ministers to resign. The paper accused the two of denying the threat from BSE in Spain.

Britain

The UK still remains the single largest source of BSE, with 1,277 cases confirmed last year. This brings the total number of cattle identified with the disease since 1987 to over 180,000.

In a related development, hundreds of haemophiliacs, who require blood-clotting agents produced from donated blood/plasma, have been warned they may have been infected with vCJD. Bio Products Laboratory confirmed that a person who donated blood in 1997 has been diagnosed with vCJD. Because of the potential risk, blood plasma for British haemophiliacs has been sourced in the USA since 1998.

The families of vCJD, or "Human BSE", victims finally look set to receive compensation, in some cases years after they have lost their loved ones to the protracted and harrowing disease. Interim payments of up to £25,000 ($37,000) could be made but with the final scale of the epidemic still an unknown factor—estimates vary from a few hundred to 250,000 —the government has been reluctant to agree to any open-ended commitment.

Elsewhere on the continent, the new EU tests have shown that the incidence of BSE in Belgium is five times higher than previous estimates, with 19 cases confirmed so far. The agriculture ministry has said incineration plants are now "saturated" and cannot process any more cattle.

The first suspected case has also been reported in Austria.

European Union

The first diagnosis of BSE was made in November 1986 at Britain's Central Veterinary Laboratory. Although the two cows concerned were from different parts of the country, they displayed the same abnormal neurological symptoms, identified as a spongy-like degeneration of the brain.

When the BSE crisis first broke out in the UK, butchers' shops on the continent put up signs saying, "no British beef sold here." European governments reacted with a mixture of nationalism, extolling the virtues and safety of their own beef, and then banning British imports.

However, a general relaxation of safety-critical standards combined with the domination of farming by massive agribusiness is not a purely British affair. This is particularly so in Europe, where the Common Agricultural Policy provides billions in subsidies to protect European farming.

The BSE crisis was both foreseeable and preventable. Its origins lie in the intensified production methods introduced in the mid 1980s, and particularly the practice of adding meat and bone meal to animal feeds. Once cattle that had succumbed to BSE were ground up and used in such high-protein food additives, a cycle was established that ensured the disease multiplied throughout the national herd.

Meat and bone meal additives have been banned in Britain since the 1990s (as were the "specified risk" materials) but continued to be exported abroad for some time. The EU has only imposed a temporary six-month ban in December last year, with no indication if it will be made permanent.

The EU Commission overhauled food safety mechanisms in the late 1990s, in the wake of the British BSE crisis but although the new procedures may bring to light more cases of BSE the Commission has no powers of enforcement. Agricultural policy is still a nationally guarded preserve. As an article in Britain's *Financial Times* newspaper noted, the "first response [by governments] to outbreaks of BSE elsewhere is not to step up their own safety inspection procedures but to launch campaigns urging consumers at home to boycott foreign beef."

There are plans to establish an EU food safety authority, but it will only be an advisory body and is not expected to come into operation for more than 10 years.

The BSE crisis is only the most critical in a long line of food safety scandals, which include salmonella and e-coli and have cost countless lives. As the BSE scandal now spreads across Europe, it confirms tragically that as long as food production and safety are subordinated to the profit system and the market, then public health will continue to suffer.