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Understanding the **BSE threat**

World Health Organization
October 2002

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Introduction

"Mad cow disease is only one of numerous new diseases that have emerged in recent years."

Changes in the way we inhabit the planet have disrupted the equilibrium of the microbial world. Microbes proliferate rapidly, mutate frequently, and adapt with relative ease to new environments and hosts. They are quick to exploit new opportunities to change and spread. Numerous factors, including those linked to human activities, can accelerate and amplify these natural phenomena, as has happened in recent years. Trends contributing to the renewed microbial threat include rapid population growth, rural–urban migration, international travel and trade, collapsing health systems, environmental manipulation, changing weather patterns, misuse of medicines, and altered agricultural and animal husbandry practices.

These changes have produced ideal conditions for human-to-human transmission of diseases, created new breeding sites for insects and other vectors that carry disease, and encouraged the emergence of antimicrobial resistance. They have also disrupted ecological systems in which pathogens and natural animal hosts have coexisted in equilibrium for centuries.

As a result, new diseases are emerging at an unprecedented rate. In the last decades of the 20th century, more than 30 new diseases – including HIV/AIDS and Ebola haemorrhagic fever – were detected for the first time in history. Bovine spongiform encephalopathy (BSE), or "mad cow disease", is one of these newly emerging diseases. Its related human form, variant Creutzfeldt-Jakob disease (vCJD), is another.

Understanding the **BSE threat**

Some questions for consumers

BSE, or "mad cow disease", is a new disease from a mysterious family of related and mostly very rare diseases. Cases in cattle were first reported in the United Kingdom in 1986. In 1996, another new disease, variant Creutzfeldt-Jakob Disease, or vCJD, was detected in humans and linked to the BSE epidemic in cattle. Consumption of contaminated meat and other food products from cattle is presumed to be the cause.

Both diseases pose many difficult scientific challenges. Answers to all questions cannot be given with absolute certainty. However, a great deal is now known about the origins of the BSE epidemic, the reasons for its spread, the tissues that are most dangerous to consume, and the likely reasons for the appearance of a related disease in humans. Most importantly, intense research, backed by practical experience, has defined a series of measures that countries can use to keep the causative agent out of the food chain and thus ensure the safety of the meat supply. When all appropriate measures to minimize human exposure are fully implemented and controlled, meat and meat-based products derived from cattle can be regarded as free from the BSE agent and thus free from any risk of causing vCJD in humans.

Here are some of the most important questions for consumers to ask their national public health and veterinary authorities. These questions are most important in countries where BSE cases have been reported. However, in view of the long incubation period and the fact that contaminated feed has been widely distributed in international trade, consumers and governments in other countries would be wise to consider these questions as well.

What are cattle being fed?

BSE is clearly linked to the practice of recycling bovine carcasses to recover so-called "meat and bone meal" protein, and then feeding this protein back to other cattle. If cattle are not being fed protein derived from the carcasses of ruminants (cattle, sheep and goats), there is virtually no risk of BSE. If ruminant protein is fed only to pigs and poultry, and if this feed has no chance to mix with and contaminate cattle feed, at feed mills or on the farm, the risk of BSE in the country is insignificant.

Does the government have a system of active surveillance for BSE?

The recent introduction of rapid screening tests, compulsory in many countries, has greatly improved the detection of cases. Such "active" detection of infected cattle, followed by their destruction, prevents entry into the feed chain of a large proportion of infectious material. News of a few cases in countries with active surveillance is more reassuring than no reported cases in countries with poor surveillance.

Are cases of BSE imported or are they being born within the country's herds?

Within cattle herds, BSE is not contagious and does not spread from animal to animal. Isolated imported cases will not spark an epidemic if the affected cattle are destroyed and the carcasses are not recycled for use in feed. Of much greater concern are cases of BSE in cattle born within the national herd, as this implies that feeding practices within the country are at fault and that many other cattle have been exposed.

Does meat come from young cattle?

The incubation period for BSE is very long: 4–5 years. During this period, cattle exposed to the BSE agent show no symptoms and, until late in the period, have no infectious material in their tissues. If cattle are slaughtered at a young age (preferably under 30 months), the likelihood that veal or beef and other bovine products can transmit vCJD is greatly reduced.

Are high-risk tissues removed and destroyed?

The agent that causes BSE is not distributed evenly throughout the animal's body, but is concentrated in certain tissues, most notably the brain and spinal cord, related to the central nervous system. Stringent slaughter practices that remove and destroy these high-risk tissues have an immediate impact on food safety and can protect consumers even when BSE is established within a country.

Are procedures in place to prevent cross-contamination in slaughterhouses?

The agent that causes BSE, and presumably vCJD, has never been detected in bovine skeletal muscle tissues, from which most quality meat is derived. However, an extremely small amount of the causative agent – less than one gram of brain (the size of a peppercorn) from diseased cattle – is sufficient to cause infection in cattle. For humans, the amount capable of causing infection is unknown but could likewise be very small. For this reason, it is vital to guard against cross-contamination. Safe slaughterhouse practices ensure that high-risk materials have no chance to come into contact with otherwise safe materials and contaminate them.

Are there any other meat products that could contain BSE?

The use of wire brushes and other mechanical tools to recover meat scraps attached to bones and the vertebral column can pull out infectious nervous tissue and contaminate meat that is otherwise safe. Such "mechanically recovered meat" is used in processed meat products. Some experts believe that the BSE agent was transmitted to humans through products containing mechanically recovered meat contaminated with nervous tissue. Techniques that prevent the inclusion of nervous tissue in mechanically recovered meat confer important protection, particularly in high-risk countries.

Are safe practices stringently controlled?

It is not sufficient to recommend safe practices. Such practices must also be rigorously enforced, ideally through legislation, and controlled through inspection by veterinary and food authorities.

Safe to eat – On the basis of current knowledge, scientists agree that some bovine products are safe, regardless of the BSE status within a given country. Bovine products considered safe to eat or use include milk and milk products, gelatin prepared exclusively from hides and skins, and collagen prepared exclusively from hides and skins. Infectivity has never been detected in skeletal muscle tissues, from which most quality meat is derived. A number of scientists believe that skeletal muscle meat is as safe to consume as milk and milk products, provided that such meat has not been contaminated during slaughterhouse procedures.

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A notorious – and mysterious – family of diseases

BSE is a transmissible brain disease of cattle characterized by progressive degeneration of the nervous system. The causative agent is unconventional and not fully understood, but is related to the agent that causes scrapie in sheep and goats. Scrapie, which is likewise not well understood, has been known for over 250 years and is reported throughout the world, with the exception of Australia, New Zealand, and some countries of South America. The transmission of scrapie to humans has never been documented.

Some scientists speculate that BSE originated when the agent that causes scrapie jumped the species barrier, through contaminated feed, to infect cattle. Others believe that a gene mutation caused the first, or "index" case. Still others cite changes in rendering practices that worked to concentrate the infectious agent. In any event, cattle are now considered to be universally susceptible to BSE.

Both BSE and scrapie belong to a notorious family of related diseases collectively known as transmissible spongiform encephalopathies (TSE). All affect the central nervous system, producing characteristic sponge-like changes in brain tissue. Recent studies suggest that, following ingestion, infection spreads from the gut to the spinal cord through the peripheral nerves, and then reaches the brain. All TSEs are incurable and invariably fatal. All can be transmitted, though the mode of transmission differs according to species and disease.

Apart from domestic ruminants (cattle, sheep, and goats), other spongiform encephalopathies have been observed in deer, elk, mink, zoo animals (wild ruminants such as nyala, gemsbok, kudu, and bison, large cats such as puma and tigers, and monkeys), domestic cats, and humans. Cases of human infection are exceedingly rare.

"BSE is a new disease from a largely mysterious family. Despite remarkable research progress, science cannot answer all questions with certainty."

The exact nature of the transmissible agent remains one of the great mysteries of modern biology. Though it can be transmitted, the agent is unlike other agents such as viruses and bacteria, and does not invoke a detectable immune response or inflammatory reaction in the host. The most widely accepted theory is that the diseases are caused by a "prion", an abnormal protein that "infects" the host by provoking the conversion of normal cellular protein to an aberrant form. The malformed protein

accumulates in the brain, where damage is expressed as a sponge-like appearance.

All TSE agents are known to be extremely resistant to inactivation procedures, including heat treatment and chemical sterilization. Some studies suggest that a TSE agent can survive dry heat for 1 hour at temperatures as high as 360 °C. In another study, prions remained active after heat treatment at 600 °C.

At present, no test exists for the detection of TSE in live animals or humans prior to the onset of symptoms. Diagnosis is based on examinations of brain tissue following death. No vaccine for prevention or curative drug is available for any of these diseases, though the ongoing quest is intense. For BSE, rapid diagnostic tests, also based on postmortem samples, have recently been introduced for use in screening animals at slaughter. Surveillance for BSE is consequently much improved. As yet, these tests are not reliably able to detect infectivity during the incubation period, except in its late stages.

"At present, no test exists for the detection of TSE in live animals or humans prior to the onset of symptoms. No vaccine for prevention or curative drug is available."

The TSEs in humans – Classical Creutzfeldt-Jakob disease (CJD) is known to exist in three forms: sporadic, familial, and acquired through medical procedures (iatrogenic). Sporadic cases have an unknown cause and occur throughout the world – even in countries where BSE is unknown – at the rate of about one per million. They account for approximately 90% of all CJD cases. Familial CJD, which is hereditary, is associated with gene mutations and makes up 5–10% of CJD cases. Iatrogenic CJD includes cases resulting from the unintentional transmission of the causative agent by contaminated neurosurgical equipment, from cornea or dura mater transplants, or from the administration of contaminated human-derived pituitary hormones. Less than 5% of CJD cases are caused by unintentional transmission through these medical treatments. Kuru is a form of TSE, different from CJD and vCJD. It was spread by ritual cannibalism, formerly practised by an ethnic group in Papua New Guinea.

Variant CJD is a new form of human TSE. It differs from classical CJD in several ways. It is probably caused by the same agent that causes BSE, the patients are usually younger (under 30 years of age on average) and their clinical symptoms are different. The duration of variant CJD is longer, and its effects on the appearance of brain tissue differ from those seen in the classical form.

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Origins of the BSE epidemic

The BSE epidemic began in the United Kingdom, where cases may have occurred as early as the 1970s. A distinctive feature of BSE is its very long incubation period, averaging 4–5 years, during which animals appear perfectly healthy. Initially, the long period during which infected cows remain healthy masked the true extent of the epidemic. Moreover, when symptoms of BSE do appear, they mimic those of several other diseases. For these reasons, BSE was not recognized as a new entity until 1986, by which time cases, many still in the symptomless incubation period, were widespread and the epidemic was poised to explode.

Recycling bovine carcasses: an established – and ultimately disastrous – practice

While the cause of the emergence of BSE remains unknown, the ultimate driving force of the epidemic has been identified. The establishment of BSE in its new bovine host and subsequent epidemic spread have been clearly linked to the use of meat and bones from cattle and other ruminant carcasses in the preparation of cattle feed.

Meat and bone meal animal feed is produced by rendering. As we now know, temperatures used during rendering are not sufficiently high to fully inactivate the BSE agent. Moreover, rendering pools wastes from the carcasses of hundreds of animals, so that if a single one is infected, the entire pool becomes contaminated. The infectious agent can thus spread far and wide. In addition, when infected wastes from cattle are fed back to other cattle, there is no species barrier to mitigate the risk of transmission.

"The driving force of the epidemic is known: the use of meat and bone meal feed contaminated with the causative agent."

Transmission of BSE occurs when cattle consume meat and bone meal feed contaminated with the causative agent. Some speculate that the epidemic began when dairy cows as calves were fed diets containing material from scrapie-infected sheep, and escalated when material from infected cows was recycled in feed and then fed back to cattle. An extremely small amount of the agent – less than one

The progression of a new disease – and of measures for its control

1986	1988		
November. BSE recognized as a new disease in cattle.	**June.** UK government makes BSE a notifiable disease. Suspected cases must be investigated and reported.	**July.** UK bans the feeding of ruminant-derived meat and bone meal (MBM) protein to ruminants.	**August.** UK introduces a compulsory slaughter and compensation scheme for detected cases.

gram of brain (the size of a peppercorn) from diseased cattle – is sufficient to cause infection in cattle.

The practice of recycling animal protein as an ingredient of animal feed, which dates back to at least the 1920s, was introduced as an inexpensive way to boost milk production and increase weight gain. Rendering continues to be regarded as an efficient way to use nutritious materials that would otherwise be wasted. The danger arises from the feeding of ruminant wastes back to ruminants. This practice went unchallenged, and apparently without consequences, for decades. When the consequences emerged, they quickly took on dramatic proportions.

"Less than one gram of brain from diseased cattle – the size of a peppercorn – is sufficient to cause infection in cattle."

Rendering: an essential public service – Rendering is a centuries-old practice that manages, economically and efficiently, an important environmental problem: the safe disposal of large quantities of animal by-products and wastes. In the rendering process, animal wastes, largely in the form of fat, bone, hide, and offal, are ground and melted down at high temperatures for a fixed time. Protein settles under a layer of fat. These products are then recovered for use as ingredients in numerous commercial products. Fat, tallow, and grease are siphoned off for use in products ranging from lubricants and lipstick to soap, candles, pharmaceuticals, ink, and cement. The heavier protein at the bottom is further processed and recycled, largely for use as a high-energy supplement in the feed rations of domestic animals.

Apart from its economic value, rendering performs an essential public service: the environmental clean-up of wastes too hazardous for disposal in conventional ways. For example, animal wastes provide ideal conditions for the growth of pathogens that infect humans as well as animals. Incineration would cause major air pollution. Landfill could lead to disease transmission. In contrast, rendering "sanitizes" the wastes. The high temperatures used are sufficient to kill almost all infectious agents – the causative agent of BSE being the notable exception. Once rendered into dry compact form, materials can be further sanitized by incineration as an additional safeguard against the spread of contamination.

The sheer quantity of wastes is another problem. Half of every cow and a third of every pig is not consumed by humans. In the European Union (EU) alone, the rendering industry handles some 9 million tonnes of animal waste each year. The recycling of such quantities is environmental clean-up at its best.

1989

July. EU bans the exportation of UK cattle born before July 1988 feed ban.

November. First case of BSE outside the UK is detected in a native herd in Ireland.

November. UK government bans the use for human food of certain high-risk specified bovine offals (SBO).

1990

April. Ban on export to EU of SBO and certain other tissues, including lymph nodes, pituitaries, and serum.

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Dynamics of the BSE epidemic

From the initial cases, detected in 1986, the epidemic spread to infect over 180 000 head of cattle in more than 35 000 herds throughout the United Kingdom (UK). When the epidemic peaked in 1992, as many as 1 000 new cases were being reported each month. The number of cases began to decline later that year, and this decline continues, now sharply. For example, in 1992 the number of confirmed cases was 36 680. This compares with 1013 confirmed cases reported in 2001.

Most cases of BSE have been detected in dairy herds, where cattle are typically older. Beef cattle are generally slaughtered before the age of 3 years. Since the incubation period averages 4–5 years, these cattle – even if infected – will not live long enough to develop the symptoms that give cause for suspecting BSE. When these "hidden" infections are factored in, experts estimate that the total number of cattle infected during the UK epidemic exceeded one million.

Moreover, infectious material concentrates in certain high-risk tissues, including the brain and spinal cord. These tissues are probably capable of transmitting BSE before the animal shows any symptoms of disease. Under artificial experimental conditions, infectivity is found some months before the onset of symptoms. Under real-life conditions, when cases actually occur on farms, the point within the incubation period when infectivity begins is not precisely known.

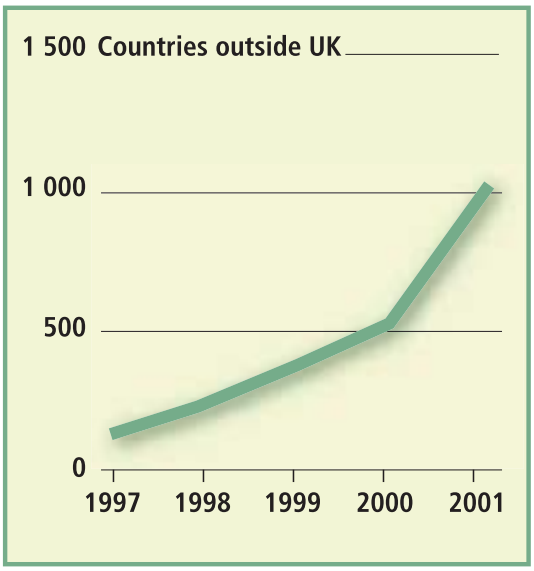
These features of the disease help to explain how large numbers of infected animals, capable of transmitting BSE, entered the UK food and feed chains during the early years of the epidemic.

"Because the incubation period averages four to five years, calves have very little infectivity."

1990			1993
May. UK establishes surveillance unit for CJD following concern about possible spread to humans.	**June.** EU bans export of live UK cattle older than 6 months.	**September.** UK bans use of SBOs from feedstuffs for all mammals and birds, including pets.	EU-funded surveillance system for CJD is introduced in 10 countries in Europe and in Australia and Canada.

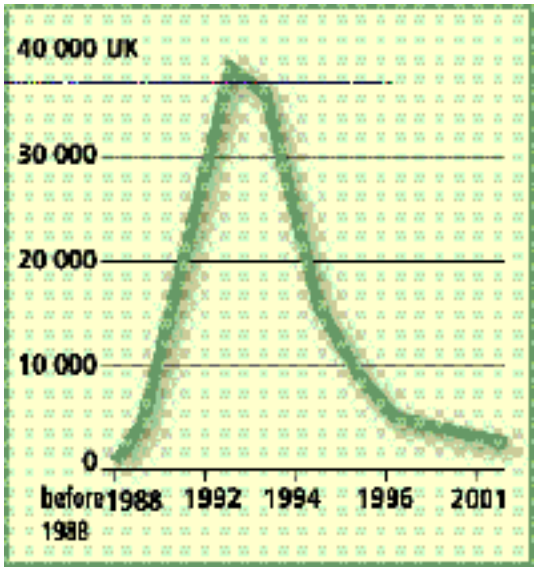
Reported cases of BSE in all countries outside of UK

Total number of cases: 2 264



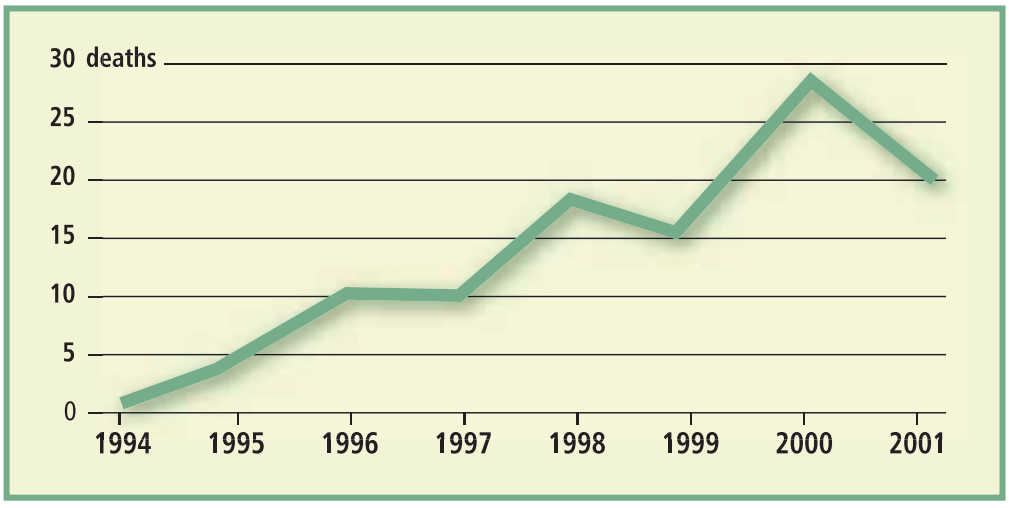
Reported cases of BSE in the UK

Total number of cases: 181 376



Global number of deaths from vCJD over time

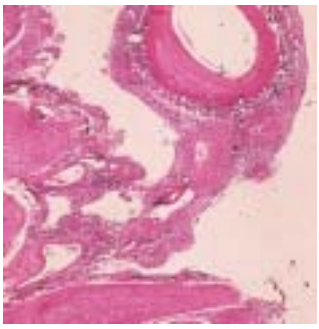
The total number is 122 deaths confirmed as caused by — or probably caused by – vCJD. Eleven additional people with a conditional diagnosis of vCJD are still alive as of July 2002



1994	1995		1996
June. The feeding of mammalian protein to ruminants is banned throughout the EU.	***July.*** EU places restrictions on the importation of beef from UK cattle.	***December.*** UK bans the use of mechanically recovered meat in human food.	***March.*** A new human TSE is identified (vCJD) and distinguished from classical CJD.

Understanding the **BSE threat**

Growing concern



A new disease in humans

Concern over the health implications of the BSE epidemic mounted considerably with the appearance, in 1996 in the United Kingdom, of 10 human cases of a new TSE similar to the well-known but extremely rare Creutzfeldt-Jakob disease. The new disease, which affects a younger age group and follows a longer clinical course, was designated variant Creutzfeldt-Jakob disease to distinguish it from the classical form.

Considerable epidemiological, neuropathological, and experimental data are consistent with the hypothesis that the agent that causes BSE in cattle also causes vCJD in humans. The most plausible route of human exposure is through the consumption of food contaminated with the BSE agent, although this has not been conclusively proven. Measures for preventing human exposure have been identified and put into effect in the UK and elsewhere, but the size of the population exposed to BSE in the UK – and possibly in other parts of the world – is not known. Materials potentially infected with BSE have been distributed throughout the world through trade in cattle and cattle products and by-products. Given the intricacies of trade in a globalized economy, the uncertainty is considerable.

"The size of the population at risk of developing the human form of BSE remains unknown."

A vulnerable world

BSE has now spread internationally, though the United Kingdom – with more than 60 times as many cases as the rest of the world combined – remains the most seriously affected country. Cases have spread beyond the original epidemic focus in two main ways: the exportation of infected cattle during the symptomless incubation period, and the exportation of animal feed based on meat and bone meal contaminated with the infectious agent.

1996

March. UK bans the use of mammalian MBM in feed for all farm animals.

April. UK introduces slaughter scheme to keep cattle older than 30 months out of food and feed chains.

June. UK introduces feed recall scheme aimed at collecting and destroying all remaining MBM.

July. Cattle passports are made mandatory for all cattle born from 1 July 1996.

As is now known, BSE is transmitted through the consumption of contaminated feed. Within cattle herds, the disease is not contagious and does not spread from one animal to another. While there is some risk that the calves of infected cows may be infected, such "vertical" transmission occurs – if at all – at a very low level and would have no significant impact on the dynamics of the epidemic.

Isolated cases occurring in imported cattle are therefore of less concern unless they were themselves rendered or recycled. Isolated imported cases have been reported in Canada, the Falkland Islands (Islas Malvinas), and Oman.

"Within herds, BSE is not contagious and does not spread from one animal to another."

BSE cases in the national herd

Far more alarming is the appearance of a case born within the national herd, as this indicates that feeding practices within the country – whether involving importation of contaminated feed or local production using recycled ruminant wastes – are the cause. Cases in native-born cattle are the most important to monitor. Since feeding practices are implicated, such cases suggest that many other cattle have been exposed, and that the disease may have been silently spreading during the long incubation period.

Outside the United Kingdom, relatively small numbers of cases of BSE have been reported in the national herds of Austria, Belgium, Denmark, Finland, France, Germany, Greece, Ireland, Italy, Liechtenstein, Luxembourg, the Netherlands, Portugal, Spain, Switzerland and, most recently, the Czech Republic, Japan, Slovakia, and Slovenia.

All but a few dozen of these "native herd" cases have been reported in six countries: France, Germany, Ireland, Portugal, Spain, and Switzerland. As with scrapie, Australia and New Zealand are considered free of BSE.

1998

January. UK prohibits specified risk materials (SRM) in feed, cosmetics, pharmaceuticals, and medical products.

2000

July. EU implements SRM ban. CJD surveillance is extended to cover 8 more countries.

2001

January. EU introduces compulsory testing of cattle and bans the use of MBM in feed for all farm animals.

December. CJD surveillance expands to include countries in Central and Eastern Europe and China.

Understanding the **BSE threat**

The special problem of emerging diseases

Most new diseases are poorly understood as they emerge. Control efforts are hampered by the lack of knowledge about the causative agent, its mode of transmission, and measures for prevention. The dynamics of the unfolding epidemic are likewise poorly understood, as are the precise factors that contribute to further spread.

Within two years of BSE being identified in the UK, the government banned the feeding of recycled ruminant-derived protein to ruminants. This action was taken shortly after investigations implicated recycled protein in the epidemic's spread. Unfortunately, it was presumed at the time that transmission occurred following the consumption of large quantities of contaminated feed. The country continued to export meat and bone meal feed with the restriction that it be fed exclusively to non-ruminants such as pigs and poultry – species that are not considered to be susceptible to BSE.

Despite this restriction, it is clear that contaminated feed was fed, in the UK and elsewhere, to cattle as well. Moreover, even in countries where meat and bone meal feed is fed exclusively to pigs and poultry, there is now a well-documented risk of cross-contamination at feedmills or even in the barnyards of individual farms. Given the very small quantity of infectious material needed to transmit the disease, the stage has clearly been set for an epidemic that can spread to other parts of the globe.

As scientific knowledge about the disease and its mode of spread progressed, a series of increasingly stringent measures was introduced and eventually proved effective in curbing the BSE epidemic in the UK. Protective measures, though inadequate at first, were rapidly modified as each gap in effectiveness was recognized. The resulting experiences have led to considerable knowledge about what fuels the epidemic, what practices place humans and animals at risk, and what works best to safeguard public and animal health. They have also demonstrated how stringent the safeguards must be in order to protect humans against the risk of acquiring vCJD.

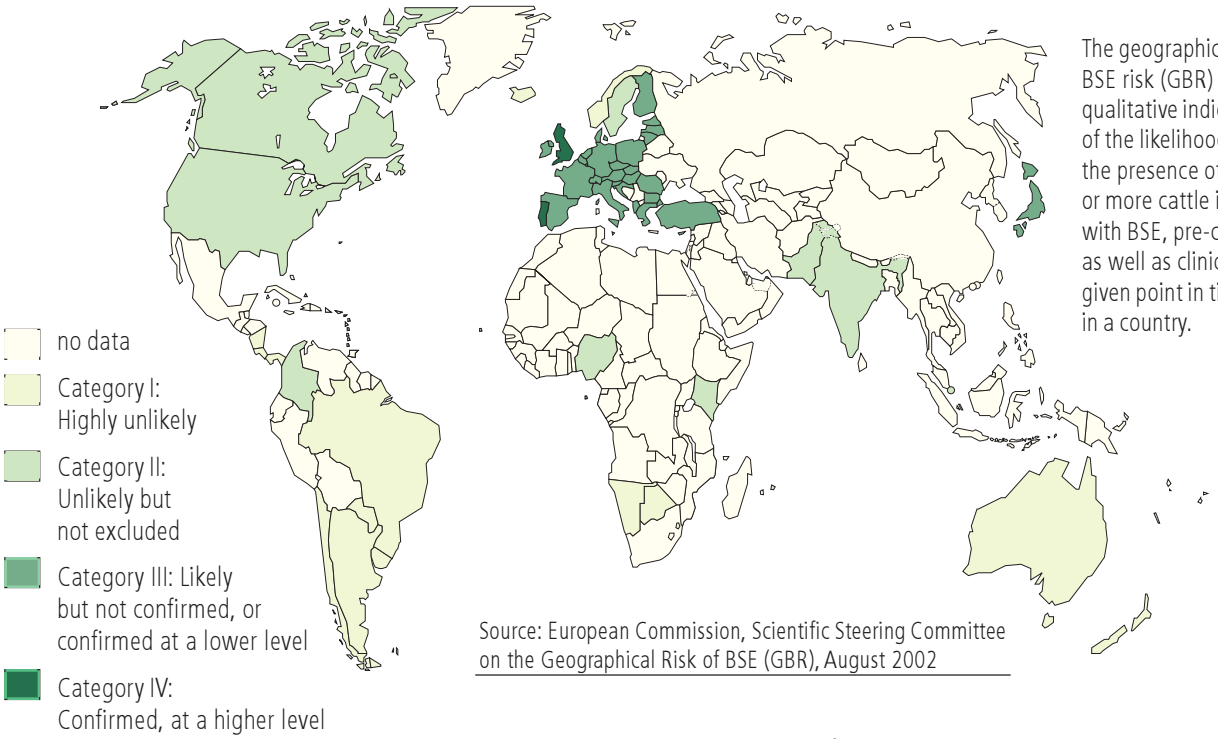
"The stage has clearly been set for an epidemic that can spread to other parts of the globe. The knowledge exists to prevent this from happpening."

Expert opinion – In response to mounting public concern and growing questions about the epidemic's future course, a high-level international technical consultation on *BSE: Public Health, Animal Health and Trade* was held in Paris in June 2001. The consultation, jointly sponsored by WHO, the Food and Agriculture Organization of the United Nations (FAO), and the Office International des Epizooties (OIE), united experts in veterinary and human medicine, leading TSE researchers, representatives of industry and consumer associations, and national regulatory authorities.

The meeting aimed to review scientific knowledge of the causes of BSE and vCJD and to identify the factors that place animals and human populations at risk. The experts also set out specific measures that can offer a strong degree of protection against these risks. Such measures are associated with both domestic feeding, rendering, and slaughter practices and the factors that determine how meat, meat preparations, bovine products, animal feed, and live animals move around the world. Though the consultation considered strategies for protecting industries from excessive trade restrictions and their repercussions, the highest priority was given to measures to ensure that infectious material does not enter the food chain at any point, especially as a consequence of international trade, and that the risk of human exposure is minimized.

By separating the facts from areas of uncertainty, the consultation produced clear advice for governments and consumers alike. A report summarizing the recommendations of the consultation can be viewed at: http://www.who.int/emc/diseases/bse/meeting.htm

Geographical BSE risk assessment



The geographical BSE risk (GBR) is a qualitative indicator of the likelihood of the presence of one or more cattle infected with BSE, pre-clinically as well as clinically, at a given point in time, in a country.

Source: European Commission, Scientific Steering Committee on the Geographical Risk of BSE (GBR), August 2002

Understanding the **BSE threat**

Preventive measures for halting the epidemic

"A global epidemic need not happen."

Though significant gaps in the scientific understanding of BSE and vCJD remain, experiences during the UK epidemic, backed by considerable scientific knowledge, provide a sound basis for devising and implementing preventive measures. Such measures can go a long way towards ensuring a safe meat supply and preventing spread of the epidemic to new countries. A global epidemic, with devastation on the scale seen in the UK, need not happen.

In defining an appropriate response, the first priority must be to protect populations from the risk of contracting vCJD. Exposure through food is by far the most important risk to consider. As no method exists to rid contaminated food of the causative agent, efforts to protect public health must rely on measures aimed at preventing the BSE agent from entering the food or feed chain at any point. When all appropriate measures to minimize human exposure are fully implemented and monitored, food can be regarded as free from the BSE agent and thus free from any risk of causing vCJD in humans.

Removing the risks

The first lesson from the UK experience is paramount: meat and bone meal of ruminant origin must not be fed back to ruminants. In countries where feed using recycled ruminant wastes is fed to non-ruminants, such as poultry and pigs, measures must be in place to ensure that this feed is not allowed to contaminate ruminant feed. Some countries have taken the further step of banning all mammalian protein-based animal feed from ruminant feeds or even from all feed for all farm animals.

The BSE agent is concentrated in certain tissues and not distributed throughout the bovine body. Measures must ensure that these infectious tissues have no opportunity to enter the food or feed chain. The brain and spinal cord account for by far the largest amount of infectivity (nearly 90%). As only a very small quantity of infectious material is believed capable of causing disease in cattle, and the infective dose for humans is unknown, all tissues suspected of containing any level of infectivity should be removed and

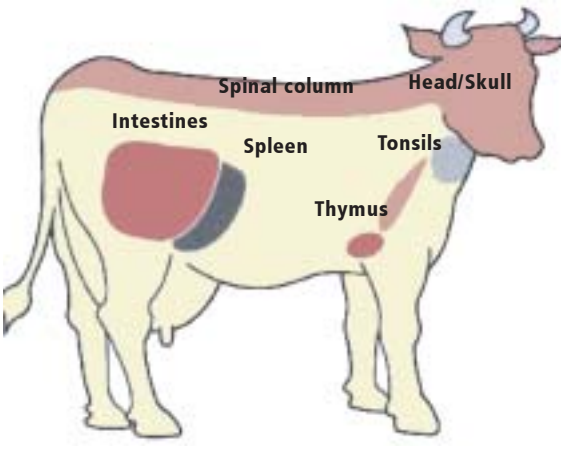
The level of risk determines the level of action

Country considered as BSE free:
may export without restriction

Country with minimal BSE risk:
may not export brains, eyes, spinal cord

Country with moderate BSE risk:
may not export brain, eyes, spinal cord, distal ileum, skull, vertebral column

Country with high BSE risk:
may not export brain, eyes, spinal cord, tonsils, thymus, spleen, intestines, distal ileum, dorsal root ganglia, trigeminal ganglia, skull, vertebral column



Source: OIE International Animal Health Code

destroyed. Use of these so-called "specified risk materials" for human food or for rendering should be strictly prohibited, as is the case in numerous countries that have introduced protective measures.

The removal at slaughter of specified risk materials also protects against the risk posed by cattle that may be incubating the disease yet do not show any symptoms and do not test positive in the new rapid tests, which are unable to detect the earliest stages of infectivity. Research shows that infectivity is also concentrated in the brain and spinal cord at the earliest stage during the incubation period when the cow becomes capable of transmitting BSE.

Mechanically recovered meat: a special risk

Bovine tissues that do not contain infectivity can nonetheless become contaminated – and endanger the food supply – following certain slaughterhouse practices. In terms of understanding all precautions needed to protect public health, one of the most important recent findings is infectivity in part of the large nerves and dorsal root ganglia that extend from the spinal cord.

This finding takes on particular significance in view of the practice of stripping muscle meat attached to bones and the vertebral column using wire brushes and other mechanical tools. In the process of producing this "mechanically recovered meat", the wire brushes can also pull out the infectious nervous tissue and contaminate the recovered meat.

Some experts believe that BSE was transmitted to humans through products such as inexpensive hamburger, sausage, meat pies, and other processed meat-based products, which contained mechanically recovered meat contaminated with nervous tissue. Since such products tend to be consumed by younger individuals, this mode of transmission might help explain why the majority of vCJD cases to date have occurred in a young age group.

"Some experts believe that BSE was transmitted to humans through processed meat products. Such products probably used mechanically recovered meat, which can be contaminated with nervous tissue."

Understanding the **BSE threat**

Active surveillance: reassurance for consumers

"Active detection of cases, followed by destruction, prevents entry into the feed chain of a large proportion of infectious material."

Much of the infectivity that could be distributed through international trade in feed, cattle, meat, and other bovine products has already been distributed. Following the introduction of control measures, materials currently exported from countries with BSE should carry very little risk provided that recommendations concerning removal and destruction of high-risk materials have been followed.

The need now is for good surveillance in all countries, especially those that may have imported contaminated feed, to monitor the dynamics of BSE as a potentially global epidemic, to introduce appropriate measures and to ensure that no new BSE foci are developing.

Until recently, surveillance depended upon reports of cases submitted by veterinarians and farmers. While useful, such "passive" surveillance detects only a portion of cases. The introduction of rapid postmortem screening tests, compulsory throughout the EU since January 2001, now makes "active" surveillance possible. Although not reliably able to detect infectivity until the late stages of the incubation period, these tests can detect infection in cattle that do not pass the stringent requirements for food animals (emergency slaughter) or that become ill and fall down while on the farm (downer cows).

In some countries, screening tests are being used on all cattle over the age of 24 months brought to slaughter. Active detection of BSE cases followed by destruction prevents entry into the feed chain of a large proportion of potentially infectious material – though not all.

Consumer concerns

When these sensitive new tests were introduced, all countries having more than a few isolated cases found more cases quickly – usually more than twice as many. Other countries, including Germany, Italy, and Spain, detected their first cases following the introduction of screening tests. Despite warnings from the scientific community that such active searching would uncover new cases, great public alarm accompanied news of these cases.

In terms of protecting consumer health, news of a few cases in countries conducting active surveillance can be more reassuring than no reported cases within countries lacking

a sound surveillance system. Outside Europe, Australia, New Zealand, and some countries of the Pacific Rim and the Americas, few countries have surveillance systems capable of reliably detecting cases.

Surveillance systems give consumers as well as trade partners the necessary confidence that potential risks are being monitored and cases can be identified. Surveillance data provide an indicator of the effectiveness of risk management measures and monitor the effect of any changes in the overall BSE risk of a region, country or zone. Countries that have not established surveillance systems, and therefore have not effectively assessed their risk, may pose a more serious hazard, to both consumers and trade partners, than countries that have detected cases and taken appropriate action.

"News of a few cases in countries with active surveillance can be more reassuring than no reported cases in countries with poor surveillance."

Safe to eat

Bovine products that are considered safe to eat or use include milk and milk products, gelatin prepared exclusively from hides and skins, and collagen prepared exclusively from hides and skins.

Infectivity has never been detected in skeletal muscle tissue, from which most quality meat is derived. A number of scientists consider skeletal muscle meat to be as safe to consume as milk and milk products, provided that such meat has not been contaminated during slaughterhouse procedures.

Keeping the BSE agent out of the food chain

- Bans on the use of meat and bone meal feed have a long-term impact aimed at eliminating BSE and the risk of vCJD entirely.
- Active surveillance, backed by an appropriate response to detected cases, likewise aims to eventually eliminate the disease, while also providing reliable information on the extent of infection – and thus the level of risk – within a country.
- Stringent slaughter practices, including the removal of high-risk tissues, have an immediate impact on food safety and can protect human populations even when BSE is established within a country.

Understanding the **BSE threat**

Interpreting the risk within countries

"As BSE does not spread from one animal to another, there is no risk that an imported case will spark an outbreak within the herd."

When a country reports BSE cases, the first question to ask is whether the case involves an imported animal or one born within the native herd. As explained earlier, a case of BSE in imported cattle is of far less concern. Since BSE does not spread from one animal to another, there is no risk that an imported case will spark an outbreak within the herd, provided that the cow is destroyed and the carcass is not recycled for use in feed.

Far more alarming is a case born within the national herd, as additional cases, caused by the same exposure to contaminated feed, will nearly always be uncovered. For cases occurring in the native herd, the number of reported cases reflects the quality of the surveillance system and tells only part of the story. More important in terms of the degree of risk are the feeding practices allowed or followed in the country.

Factors that influence risks

Altogether, the risk to humans and animals can be assessed by considering several factors.

- If cattle are not being fed ruminant protein, there is virtually no risk of BSE.
- If meat-and-bone-meal feed is used only for pigs and poultry, and no cross-contamination occurs, the risk of BSE is insignificant.
- If a country produces meat-and-bone-meal feed in strict accordance with recommended measures (no use of high-risk tissues, adequate heat treatment), the risk of BSE is reduced.
- If specified risk materials are diligently removed and destroyed, the risk to human health is likewise reduced.
- The risk to human health is further reduced when safer slaughter practices protect against contamination.
- If cows are slaughtered at a young age, the risk of infectivity is reduced.
- Precautions to prevent the contamination of mechanically recovered meat with nervous tissue confer an additional level of protection against human exposure.

Taking a serious threat seriously

Of all the lessons learned from the BSE epidemic in the UK, one in particular stands out: BSE is a threat that must be taken seriously by all. As the disease emerged, what might have been an isolated incident escalated into a catastrophe partly because recommendations and restrictions were not strictly followed either in the UK or in importing countries.

Countries with no detected cases of BSE should not become complacent in the face of a potential global epidemic. The extremely low initial incidence and the low within-herd incidence of BSE cases, long incubation period, and non-specific nature of the early clinical signs can delay the detection of the first cases of disease and mask epidemic spread. As the WHO/FAO/OIE consultation underscored, countries should also be aware that their trading status may be dependent upon conducting a risk assessment for BSE and subsequently introducing appropriate preventive measures.

Shared responsibility – from farm to fork

The measures needed to prevent a global epidemic of BSE are now well defined, backed by considerable scientific knowledge as well as practical experience, and feasible to implement. Prevention is a responsibility shared by all those involved in the food and feed chains – from farm to fork. All those with a role to play – from farmers and feed producers to abattoir workers, butchers, veterinarians, inspectors, and government authorities – must accept the serious consequences of their failure to adhere to preventive measures. As experience has so clearly demonstrated, the stakes are extremely high in terms of lost livelihoods, damaged industries, reduced trade, weakened national economies, and immense human suffering from a devastating disease.

"Prevention is a responsibility shared by all those involved in the food and feed chains – from farm to fork."