

Commonly asked questions about BSE
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1. What is Bovine Spongiform Encephalopathy (B.S.E., also known to the media as “mad cow disease”)?

This is an unusual disease of cattle that causes degeneration of the brain. In fact the name really tells you all you need to know:

- Bovine – affects cattle
- Spongiform – sponge like
- Encephalopathy – brain disease

Essentially the disease causes brain cells to die. When they die they leave behind microscopic holes. When the brain is examined under a microscope the holes can be easily seen and the brain looks a little like a sponge. The damage to the brain results in changes in the animal’s behavior and also cause problems with walking.

2. What is a prion?

BSE is one of a group of related diseases (known as Transmissible Spongiform Encephalopathies (TSEs)) that are seen in a number of species including;

- Scrapie – Sheep
- Chronic wasting Disease (CWD) – Deer and Elk
- Transmissible Mink Encephalopathy – Mink
- Creutzfeld-Jacob Disease (CJD), GSS, FFI, Kuru – Humans

These disease show a number of similarities; the time from infection to disease is very long (typically measured in years) and the agent that causes the disease is incredibly hard to destroy (much, much harder than either the bacteria or viruses that causes most infectious disease). Furthermore despite years of research scientists were unable to find an infectious agent. In the mid 1980s an American research had a theory that scientists had been looking in the wrong place, perhaps there was no virus or bacteria involved. This led to the theory that pure proteins could be infectious (hence Prion – proteinaceous infectious particle).

We now know that the prion protein is a natural protein found in the brains of most animals (we don’t know what it does yet). Like all proteins it has a very characteristic shape, which is essentially a series of spirals. When an animal develops a spongiform encephalopathy the protein essentially re-folds into a different shape, essentially a flat sheet. The significance of this change is that the protein is incredibly tough in this new shape and cannot be destroyed by the cell’s natural recycling processes. The abnormal proteins accumulate into long strands (fibrils) and eventually kill the cell.

We now think that if an animal eats abnormal prion protein, the protein is able to survive digestion, get out of the gut and make its way to the animal’s brain where it able to induce the natural prion protein to change shape and the process then starts a chain reaction with more and more prion protein being produced.

3. What does BSE look like?

As BSE affects the brain the two main signs of the disease are changes in the animal’s behavior and changes in the way that the animal walks. It is important to recognize that on a worldwide basis almost all cases of BSE have been found in dairy cattle. This is important because obviously dairy cattle are much more commonly handled and producers tend to know their animals very well. Consequently dairy producers can often recognize very subtle changes in behavior that would go unmissed on a beef farm.

The main signs include:

- Increased nervousness: Animals are typically reluctant to pass through doorways and become increasing agitated when approached, there is excessive movement of the ears, licking of the muzzle and a fine tremor may be noticed.
- Animals will also respond in a more violent manner to sudden noises, changes in light or sudden stimulation e.g. touching the legs with a broom handle.
- The increased nervousness may also manifest as an increase in aggressive behavior towards humans and other cattle. A commonly reported sign in the UK was increased kicking when the cow was milked.
- The earliest changes in the animal’s ability to walk are an increased sway to the hips as she walks. This will slowly progress to an obviously peculiar gait. The animal will slip and fall more frequently until over a period of several months she eventually becomes a “downer”.

4. Where did BSE come from?

The original source of BSE is not known. The first case was diagnosed in the UK in 1986. It is likely that the farm on which the first case was seen had seen two previous cases in 1985. Based upon statistical modeling epidemiologists think that the first case of BSE likely occurred some time in the late 1970s. Three possible sources of BSE have been recognized:

- BSE may have come from scrapie in sheep following the feeding of contaminated rendered Meat and Bone Meal (MBM). This theory was the original theory that the British government used to explain the discovery of BSE. However, extensive testing of the BSE agent has shown that it is different from all the forms of scrapie previously identified.
- The second possible theory is that BSE is naturally a disease of some form of exotic ungulate such as African antelope. A number of these animals were present in Britain in the public zoos. When an animal died the carcass was rendered to MBM.
- The final theory is that BSE may be an extremely rare natural disease of cattle (similar to CJD in humans, which occurs spontaneously at the rate of about 1 case/ million). This natural disease was unfortunately greatly multiplied by the feeding practices used in Great Britain during the 1980s.

5. How did BSE spread?

We are now fairly confident that the major method of spreading BSE was the practice of using rendered MBM to feed dairy cattle (especially dairy calves) in the UK. Rendering essentially involves taking waste tissues from carcasses and cooking them at high temperatures and pressures. This essentially produces three products:

- Water
- Tallow - fat
- Meat and Bone Meal – protein

Prior to the discovery of BSE rendering was thought to destroy all infectious organisms. We now know that under certain conditions it is possible for the BSE agent to survive the rendering process. Hence, if an animal with BSE dies and is rendered the MBM produced still contains the infectious prion. If this material is fed to another calf that animal may become infected with BSE. The feeding of MBM to cattle was a widespread practice in the UK dairy industry, this was because it was freely available and was a cost effective source of protein. Furthermore other sources of protein used in Canada are not available in the UK due to the climate e.g. Alfalfa and Soya bean

6. How did BSE come to Canada?

It is now thought that BSE may have come to Canada from the UK. Following the discovery of BSE and the start of the UK epidemic Canada banned the import of cattle and cattle products from the UK in 1990. Between 1982 and 1990 182 cattle were imported from the UK.

One of these animals developed BSE in 1993 and was culled. Following this event the CFIA (Canadian Food Inspection Agency) started a tracing program to identify the imported British cattle. 113 of the animals were still alive and were destroyed. 68 animals had previously died or been slaughtered. A detailed check of the records show that 10 of these 68 animals originated from farms in the UK that had subsequently developed at least one case of BSE. These 10 animals are therefore considered at being at risk of having carried BSE. In fact 2 of the animals originated from the same farm as the animal that developed BSE. Since they were of a similar age there is a strong probability that these animals may have been exposed to BSE.

Currently there is an extremely detailed investigation trying to determine where these animals either died or were slaughtered and what happened to the offals after death. The goal of this investigation is to determine if there is a geographical area at increased risk for BSE within Canada.

7. Why has BSE hit Canada so hard?

The simple answer to this question is that Canada was a large exporter of beef. The discovery of a home raised case of BSE led many countries to ban the importation of Canadian beef. There really is no scientific reason for the complete importation ban. The protocols put in place by the CFIA in Canada are adequate to protect both human and animal health and our beef does not represent a risk to human health (see below). Furthermore the OIE (World Animal Health Organization) has guidelines in place to control the trade between countries after the discovery of BSE. These guidelines specifically allow the export of beef from young animals providing that certain criteria are met. Canada meets all these criteria.

8. What is the risk to human health?

Prior to 1996 the position of the British government was that since other spongiform encephalopathies did not spread to humans it was extremely unlikely that BSE posed a risk to human health. In fact the British government repeatedly stated that “beef was safe”.

In 1995 a new form of CJD was identified in Britain. The disease differed from the classical form of CJD in that it mainly affected young people and the symptoms and pattern of the disease was also different. It was named New Variant CJD (vCJD). Laboratory research in subsequent months linked this disease to BSE.

To date there have been approximately 150 cases of vCJD worldwide (140 of them in the UK, the others in people who had lived in the UK). When the disease was first discovered there were predictions that these disease would result in 100,000s of deaths. That has clearly not been the case and current estimates are that there will be very few additional cases.

To put things in perspective, BSE was present in the UK for about 10 years prior to its discovery. During that time period many affected animals were slaughtered and entered the human food chain. At that time almost the entire carcass was being used for human consumption, including the tissue with high levels of infectivity such as the brain and the spinal cord. Britain has a population of approximately 60 million. There have been very few cases of vCJD when you consider that probably the entire population was exposed to the disease at some point!

In Canada since the early 1990s brain and spinal cord has been routinely removed and does not enter the human food chain. Following the discovery of BSE in Alberta in 2003 these regulations have been further tightened.

In comparison to the UK, the level of BSE in the Canadian cattle population is extremely low. The high risk materials are removed and the risk of transmission is extremely low anyway. Furthermore, infectivity cannot be found in experimentally infected animals until they reach at least 18 months of age. Consequently the feedlot animals that we slaughter for high quality steak products do not contain infectious material.

The risk of transmission of BSE from cattle to humans is so low in Canada as to be non-existent.

9. Why does BSE cause so much hysteria?

This is a difficult question to answer. I think part of the problem comes from the fact that from the outset the British media coined the term “mad cow disease” which is obviously highly emotive. Next, the original outbreak in the UK was poorly managed. The British government stuck so strongly to the “Beef is safe” point of view that when the risks of vCJD became known there was a public backlash of concern.

Finally, BSE has become a highly political issue. Governments can use BSE to put up trade barriers. Whenever a government uses BSE as a trade barrier, this sends a message to their population that BSE is “dangerous” and further amplifies the public’s concerns about the disease. Such a strategy is not without risk, if you convince your population that BSE is dangerous and then discover BSE within your own country you will see a massive public over reaction to the disease as was seen in Germany.

Our best defense in dealing with BSE is public education about the disease and also about the farming industry as a whole.

10. What happened in the UK?

More than 95% of cases of BSE have occurred in the UK. The reason that the outbreak was large was mainly due to the fact that the disease spread unnoticed in the British cattle herd for up to 10 years. With affected animals dying and being rendered and feed back to other cattle through the widespread use of MBM in dairy cow rations. Currently, there have been more than 180,000 cases of BSE in the UK, although the epidemic has slowed considerably and now appears to be almost over. The cessation of the epidemic is credited to the implementation of rigorous feed bans to prevent potentially infected material being fed back to other cattle.

No other country has had anywhere near the problems with BSE. Most of the countries affected with BSE had very close trading ties with the UK e.g. Southern Ireland, France, Portugal.

11. What countries have been affected by BSE? (Shown below: Country and Number of Cases)

Austria	1	Greece	1	Poland	9
Belgium	121	Ireland	1325	Portugal	845
Canada	2	Israel	1	Slovakia	12
Czech Republic	8	Italy	88	Slovenia	3
Denmark	13	Japan	9	Spain	378
Finland	1	Liechtenstein	2	Switzerland	451
France	849	Luxembourg	2	United States	1
Germany	295	Netherlands	70		

12. What has Canada done about BSE?

Canada had learnt a lot from the British experience of BSE.

Firstly, the high risk materials have been removed from the human food chain since the early 1990s.

Secondly, in accordance with the recommendations of the OIE, Canada has had a BSE surveillance program in place since the early 1990s. Animals deemed to be at risk of having BSE are routinely sampled and checked for evidence of the disease.

Secondly in 1997, the feeding of MBM of ruminant origin to ruminants was banned effectively breaking the cycle of infection.

Finally the Cattle identification program was introduced making the tracing of animals much simpler in the event of an investigation into cases of BSE.

It is likely that further feeding bans are likely to come into force preventing the use of SRMs (Specified Risk Materials – the tissues of the carcass recognized as carrying infectivity for BSE including the brain and spinal cord) in the rendering process or completely banning the use of mammalian MBM in animal feed.

13. How do you diagnose BSE?

BSE can only be diagnosed by examination of the brain from a dead animal. Initially the diagnosis was made by making microscope slides from the brain and examining them under the microscope. This process is extremely time consuming. It takes at least 7 days to make the slides and only a specially trained veterinary pathologist can make the diagnosis.

A number of so called rapid test are now available. All of these test work on the principle that a specific portion of the brain can be essentially liquidized and then digested with enzymes. The brain tissue is fully removed and only the very tough abnormal prions remain. The prions can be detected by a number of assays.

Many research groups around the world are working on a simple blood test for BSE. This is proving extremely difficult to develop and although it is likely that such a test will become available in the near future we do not have such a test as yet.

14. Will more cases of BSE be found in Canada?

Unfortunately, our experience with the disease in other countries leads us to suspect that there will be additional cases diagnosed over the next few years. It is likely that the number of cases will be low and because of the policies already in place they will not have any impact on human health. In fact one could go so far as to say the fact that we will be finding these cases is evidence that our surveillance system is working!

Our concern should actually be focused on how will other countries respond to these new cases? This unfortunately is an unknown, we can only hope that through education and a willingness to base policy on scientific evidence they will respond in an appropriate and logical manner.