Bovine spongiform encephalopathy in Switzerland – the past and the present

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Summary
The first case of bovine spongiform encephalopathy (BSE) in Switzerland was diagnosed in November 1990, although the risk factors identified in the United Kingdom were not all present in Switzerland. At 29 December 1998, a total of 282 BSE cases (all animals born in Switzerland), had been recorded. The number of cases is declining, with fourteen cases diagnosed in 1998 (at 29 December 1998).

The measures taken are effective. However, the potential weakness in current activities that allow the disease to exist are being evaluated. Further measures to avoid even minimal risk for human and animal health will be taken if required. Experience gained from the past indicates that it is necessary to review the situation constantly and to inform the public and professionals regularly and in a transparent manner.

Keywords

Introduction
Bovine spongiform encephalopathy (BSE) belongs to a group of related diseases known as transmissible spongiform encephalopathies (TSEs). The group includes scrapie in sheep and goats, transmissible mink encephalopathy, chronic wasting disease in mule deer and elk, and human TSEs, such as Creutzfeldt-Jakob disease, kuru and others.

All forms of TSE typically exhibit a slow degenerative progress of the disease that affects the central nervous system with characteristic spongiform lesions and abnormal prion protein (PrP\textsuperscript{Sc}; prion protein scrapie) in the brain of the affected animals.

Clinical signs of BSE are mainly behavioural changes which include a nervous appearance, alterations in normal posture and movement, and loss of weight (11, 20, 38). The average time between infection and the onset of clinical signs is 5 years. All affected animals die, and there is no treatment or vaccination option (36). Although clinical signs of BSE are fairly typical, no diagnostic assay of live animals is available for either the aetiological agent or the by-products of infection. Confirmation of a diagnosis of BSE is made by post-mortem examination of brain tissue. A clinical diagnosis can also be confirmed by immunocytochemical detection of PrP\textsuperscript{Sc}.

Bovine spongiform encephalopathy was first described in the United Kingdom (UK) in November 1986 (35). Based on current knowledge, the infection was transmitted by the ingestion of insufficiently rendered meat-and-bone meal (MBM) which contained infectious organs of infected animals (36, 37).

Meat-and-bone meal supplements contaminated with a scrapie-like agent, originally from sheep, but later from cattle, were added to feed concentrates (36, 37). The major risk factors identified as being important for the occurrence of the BSE epidemic in the UK are as follows: a large sheep population (relative to that of cattle), the presence of scrapie in the sheep population, the inclusion of ruminant-derived protein in cattle feedstuffs and an ineffective rendering process.

The first BSE cases outside the UK were diagnosed in the Falkland Islands and Oman in 1989 in animals imported from the UK. The first cases in cattle that were born and raised in countries outside the UK were reported in the Republic of
Ireland in 1989 and in Switzerland in 1990. Other countries with native cases of BSE were Belgium, France, Luxembourg, the Netherlands and Portugal. Imported cases have been diagnosed in Canada, Denmark, the Falkland Islands, Germany, Italy and Oman (29).

The purpose of this paper is to describe the occurrence of BSE in Switzerland and the actions taken to avoid further infection and to eradicate the disease.

The paper is structured as follows: description of the occurrence of BSE and the measures taken before the detection of the first case; the additional action taken after the first BSE case; the current BSE situation in Switzerland and the additional measures implemented after the declaration of the BSE-variant-Creutzfeldt-Jakob disease (V-CJD) connection in the UK (40).

Switzerland prior to the occurrence of bovine spongiform encephalopathy

Risk analysis for potential presence of bovine spongiform encephalopathy in Switzerland

Between 1988 and 1989 detailed data about the new disease and the measures taken were received from the UK. Much information was available after the first ad hoc meeting on BSE held at the Office International des Epizooties (OIE) on 28 and 29 September 1990 (28). The information received was evaluated and a decision was made to review the potential risk factors for BSE in Switzerland. The risk factors identified in the UK were not all present in Switzerland.

In contrast to the UK, scrapie is very rare in Switzerland; before 1991 only one suspected case had been diagnosed in 1982 in a goat (14).

Switzerland has a relatively small sheep population in relation to the cattle population. The cattle population totalled 1,830,000 whereas the figure for sheep was 409,400 according to the 1991 statistics (12).

The rendering system in use was quite efficient. In contrast to the UK where a reduction of the temperature of the sterilisation process of MBM production was introduced in the early 1980s, until 1993 Swiss animal waste-derived protein was processed at 120°C for 30 minutes or 130°C for 20 minutes with wet heat and pressure. Since then, waste must be processed at 133°C for 20 minutes at 3 atmospheres (4).

The only risk factor present was the use of ruminant-derived proteins in cattle feedstuffs (21). Until 1 December 1990, the Swiss dairy cattle population was exposed to animal-waste-derived proteins which had been legally mixed with feed concentrates to produce feedstuffs. The concentrates usually contained approximately 2.6% MBM. Some calf feed concentrates, however, might have contained up to 7% MBM.

Another important risk for countries outside the UK was obviously the importation of MBM and live cattle from the UK. From 1985 to 1990 Switzerland imported an average of 12,664 tonnes of animal-waste-derived protein for feeding purposes each year from various countries. However, only a negligible 12 tonnes of MBM were imported directly from the UK according to British export statistics (21, 22). Also considered as a low risk factor was the importation of live animals from the UK. Before 1990, an average of only 5,000 live slaughter cattle per year were imported, mainly from France (27).

Establishment of a surveillance system

The decision made after reviewing the information was to establish a BSE surveillance system in Switzerland with the objective of demonstrating the absence of the disease.

An essential component of a surveillance system is the role played by farmers and veterinarians who need to be informed about the disease, in addition to a laboratory which is capable of performing diagnosis.

An information campaign was launched in 1989 to familiarise veterinarians with the typical symptoms of BSE; information about the typical clinical signs of BSE was frequently presented as articles in journals for veterinarians, farmers and other professionals, and in presentations. Veterinarians were invited to send all cattle with neurological disorders for specialised pathological examination. In the same year, a reference laboratory was set up and research on diagnostic methods was promoted. The designated institute, the Institute for Animal Neurology at the Veterinary Faculty of Bern, had been studying neurological diseases in all animal species for nearly 60 years. In the 1970s this laboratory initiated studies to determine the occurrence and frequency of neurological diseases, such as rabies, among animals in Switzerland (15). Research workers were aware of the occurrence and relative frequency of neurological diseases in farm animals in Switzerland and, furthermore, received training in diagnostic methods for BSE in the UK.

Preventive measures

Although the foreign trade statistics indicated that hardly any MBM had been imported directly from the UK into Switzerland, the Swiss Federal Veterinary Office recognised the potential danger and stopped granting permits for importing such material.
On 13 June 1990 the Swiss Federal Veterinary Office formally implemented an import ban on live cattle, MBM, and other related products from the UK.

In summary, the risk of BSE occurring in Switzerland seemed negligible.

Switzerland after the occurrence of bovine spongiform encephalopathy

Detection of the first case
The first case of BSE in Switzerland was diagnosed in November 1990 (13). The cow concerned was slaughtered in July 1990 after demonstrating central nervous symptoms that were suspect of rabies or listeriosis. In compliance with the initiated routine to send all brain material from cattle with neurological symptoms to the Institute of Animal Neurology, the brain was examined at the Institute in October. For confirmation, the material was sent to the UK. The definitive result was confirmed in the beginning of November 1990.

Action taken after the first case
After the detection of the first case of BSE, it seemed obvious that this would not be an isolated incident. A short time later, on 4 January 1991, the second case was diagnosed.

In November and December 1990, Switzerland implemented the first regulations to control BSE, similar to those enforced in the UK and based on the recommendations of the OIE. Strict precautions were taken to exclude any potential risk to consumers of bovine products and to prevent the importation, recycling and feeding of products potentially containing the BSE agent.

Measures were taken to both eradicate BSE and to avoid the transmission to other species, including humans. Although no evidence of transmission of the BSE agent to humans existed at that time, the main reason for these measures was to protect consumers. Measures were implemented against scrapie simultaneously.

Surveillance and other measures on the farm
The 'Ordinance on immediate measures against spongiform encephalopathies in ruminants' (3) came into force on 1 December 1990. With the implementation of the first measures, BSE and scrapie notification were made mandatory. This notification mandate was essential for the detection system. Efficient notification of all suspect cases was guaranteed by full compensation of the value of the animal and of all the costs related to the management of the case, such as the diagnosis, visits by the veterinary practitioner, incineration and others. All suspected animals were to be slaughtered and the brains examined. Additionally, semen, non-fertilised oocytes and embryos from BSE-affected animals were to be disposed of.

Once aware of these first cases, the farmers and veterinarians were alarmed and more motivated to identify suspect cases. Benefiting from the early information campaign, veterinarians and farmers were already aware of the typical symptoms of BSE and therefore had no problem in identifying suspect animals. By 1990, more than 20 suspect cases had been sent to the reference laboratory. In 1991, 113 cases were identified as suspects, of which only 8% were confirmed as BSE (Fig. 1). At the beginning, the percentage of confirmed cases was low relative to submissions. Practitioners were indiscriminate in their submission of neurological cases as they were unfamiliar with the typical clinical signs of BSE. After 1995, the number of suspect cases declined because there were fewer BSE cases and practitioners were becoming more experienced in the diagnosis and treatment of neurological diseases.

![Diagram](image-url)

**Fig. 1**
Bovine spongiform encephalopathy (BSE) in Switzerland: cases investigated between 1991 and 1998

After the occurrence of the first case of BSE, the existing surveillance system was intensified and a second laboratory was designated. An efficient surveillance system with two diagnostic laboratories has been in operation since 1990.

An efficient epidemiological reporting system for BSE cases was implemented; the district (cantonal) veterinarians collect information on BSE-affected farms using a special reporting form and checklist, all available information is then entered into a central database and is regularly analysed at the Swiss Federal Veterinary Office.

Surveillance should not be restricted to BSE alone, but extended to the other TSEs as well. In response to the need for BSE surveillance, a special unit was created in 1994 at the Institute for Animal Neurology and was called the 'Neurocenter', an indication that it was to encompass all neurological disturbances in animals, not only those linked to cases of BSE. It is not sufficient to have a unit dedicated to the identification of neurological disorders; the unit must also be...
able to assess the sensitivity of the methods used. When the unit in Switzerland started to report brain lesions such as tumours, which are even rarer than the low incidence of spongiform encephalopathy, it soon became evident that the methods used were highly sensitive.

Measures concerning feed
Epidemiological investigations in the UK indicated that BSE was caused by the consumption of infected feed, probably due to the inclusion in cattle feed of protein derived from scrapie-infected sheep (36, 37). Thus, it was clear that the key requirement was to eliminate the possibility that cattle might be exposed to the agent through feed. The most important measure to prevent the transmission of the BSE agent to cattle is the ban on the use of mammalian MBM in the production of feedstuffs for ruminants, the sale as feed and the feeding of MBM to all ruminants. To determine that MBM is not included in cattle feedstuffs, a number of samples from feedmills are checked microscopically each year by the responsible authority, the Swiss Federal Research Station for Animal Production. Only one sample with a considerable amount of MBM was found in 1996; legal action was taken. A small number of samples with slight MBM-contamination was detected, which was probably due to cross-contamination through pig or poultry feed. The effect of the feed ban was clear (Fig. 2); however, BSE has occurred in animals born after the ban.

Measures concerning human health
There has been no evidence of the transmissibility of BSE to humans. Nevertheless, on 8 November 1990 the use in food of the brain, and since April 1991 the entire skull, eyes, spinal cord, spleen, thymus, intestines, visible lymph and nerve tissue and lymph-nodes was banned from cattle over the age of 6 months (1). The judgement of the potential contamination of the organs was mainly based on scrapie experiments (17, 18); at the time only limited results from BSE infectivity studies were available.

Additionally, all carcasses of affected animals were to be incinerated. The ban on marketing milk from BSE-suspected or -affected cows was introduced at a time when there was no evidence that milk was not a risk. Current studies on the risk in milk are available and none show a risk of transmitting the disease (32, 41). However, the consumption of milk from BSE suspects should still be avoided as a precautionary measure. Furthermore, in 1994 the Swiss Federal Veterinary Office conducted a risk analysis to determine the potential risk of transmission of the infectious agent of BSE to slaughter-house workers. The study concluded that the risk to these workers in Switzerland was very small and no additional measures were necessary (24).

Import measures
The import ban on live cattle, feed containing material of bovine origin and meat and meat products from the UK was already in effect in June 1990 (2).

Since January 1991, the importation of MBM from countries other than the UK is only possible under certain conditions.

Epidemiological analysis
As of 29 December 1998, a total of 282 BSE cases were recorded, all of these animals were born in Switzerland. Until 1994, the number of cases doubled from one year to the next. In 1995, when the epidemic curve reached a peak with 68 cases, the increase in BSE cases was markedly lower than in previous years. In 1996, the number of cases was 45, which corresponds to a reduction of 33% compared with the previous year. The decrease continued in 1997 and 1998 (Fig. 3). Fourteen cases have been diagnosed in 1998 (as of 29 December 1998).

In 97% of the affected herds, only one BSE case was observed. Two cases have been diagnosed on seven farms and three cases on one farm. Forty-five of the affected animals (as of 29 December 1998) were born after the ban on feeding MBM to ruminants, which was introduced on 1 December 1990.

Only adult cows in dairy herds were affected. No male animal has been involved.

Geographical distribution
The first cases of BSE were found in several parts of the country and widespread distribution has been maintained.
ever since. No cases were reported in only three cantons, which were all mainly urban areas. Approximately two-thirds of the cases were born and raised on the same farm, one third were purchased. Most cases were diagnosed in the cantons with the highest cattle population.

Age
The average age of the affected animals at the time of slaughter was 5.4 years. About 70% were between 4 and 5 years old. The youngest affected animal was 32 months, the oldest 10 years old (Fig. 4).

![Age distribution of cases of bovine spongiform encephalopathy in Switzerland](image)

*Fig. 4 Age distribution of cases of bovine spongiform encephalopathy in Switzerland*

Breed
Four breeds were represented among the BSE cases. Simmental and Red Holstein and their crossbreeds (45%) and Swiss Brown (45%) were the most frequent. These are the two major dairy breeds of Switzerland. The two minor breeds affected were the Holstein and Eringer.

Source of the disease in Switzerland
Epidemiological investigations suggested that the occurrence of BSE in Switzerland could most probably be traced back to the indirect import of potentially contaminated feed compounds from the UK. Meat-and-bone meal was mainly imported from neighbouring countries especially France, Benelux and Germany, which imported MBM from the UK (22).

A retrospective case-control study was conducted to investigate the major risk factors for the occurrence of BSE at both farm and individual levels with particular emphasis on feeding with animal-waste-derived products (26). The results excluded a possible association with risk factors other than feed. Some of the factors studied were the farm features, the herd health and reproduction management, and the feeding management.

The cases born after the feed ban
The first BSE case, born after the feed ban (BAB) was diagnosed on 8 September 1993. The animal was born in January 1991 and at 32 months was the youngest animal affected in Switzerland. The next BAB cases were not diagnosed until 1995 and were all born in December 1990 or in 1991. The first BAB cases were not very alarming because the explanation was that the farmers still used feed containing MBM in 1991, which was in stock on the farm. The first BAB cases born in 1992 and 1993 were diagnosed in 1996. Forty of the affected animals were born after the feed ban (1 December 1990) including four cases born in 1994 (Fig. 2).

All the BAB cases have been investigated with a special questionnaire asking for feeding practices and possible factors of vertical and horizontal transmission (19, 23). Until now no risk factor other than feed has been identified. The questionnaire included questions for evidence of vertical transmission, iatrogenic transmission or other methods of transmission. There was no evidence of vertical transmission in any of the cases. None of the mothers of the BAB cases still alive presented any sign of neurological disease and no evidence for two or more BABs with a common dam was found. The most probable explanation is a cross-contamination of cattle feed through pig or poultry feed containing MBM. The production of pig or poultry feed which legally may contain MBM and the subsequent production of ruminant feeds in the same feed mixing plant could lead to cross-contamination (34); the same could have occurred during transport. In only three cases was there evidence that the farmer was feeding pig or poultry feed to the cattle by intention.

Progeny of BSE-affected cattle have been recorded, particularly through the tattooing of the letters 'BSE' in the left ear. Moreover, export of these animals has been prohibited. In the autumn of 1996, on the basis of the British study on the possibility of vertical transmission, a campaign to slaughter all direct offspring of BSE-confirmed cows was initiated (5). About 180 culled animals were examined within a research project (10, 16). They were examined clinically before slaughter, and tissue samples were recovered for further investigation. Brain tissue from all animals was examined pathohistologically and immunocytochemically. No spongiform lesions of BSE and no abnormal PrP were detected.

Additional measures
In 1996 and 1998, additional measures were implemented, based on new scientific results and the analysis of the current BSE situation in Switzerland.

Measures on the farm
Since December 1996 it has been mandated that if a BSE-affected animal was born before 1 December 1990, all bovines born before 1 December 1990 on the same farm must be destroyed. If the affected animal was born after 1 December 1990 (date of implementation of the feed ban) all bovines on that farm must be destroyed. The carcasses must be disposed of as animal waste. If the affected animal was not
born and raised on the same farm, analogous measures are conducted in the herd of origin of the case (6).

Farmers were compensated at a rate of 90% of the commercial value for the animals which were destroyed. After disposal of the animals the farms are declared 'free of BSE'.

Most of the culled animals were examined clinically, histologically and immunohistochemically (9, 33). Of a total of 1,761 animals tested, 6 showed no clinical signs but did show an accumulation of the infectious PrP in the brain; three of these animals also presented spongiform changes in the brain.

Since July 1998 any animals from BSE-affected farms, which have been fed with the same feedstuffs as the BSE-affected animal and which have been sold to other farms have to be marked and registered.

Measures concerning feed

Since May 1996 the use in feedstuffs of brain (within the skull), spinal cord and eyes from cows has been banned – these organs have to be incinerated (5). All carcasses of BSE-affected animals and of dead animals of any species have to be disposed of by incineration. With this measure, the risk of cross-contamination of cattle feed with the infectious agent during production or transport has been minimised. Additionally, the risk of infection through feeding of pig or poultry feed, intentionally or not, to ruminants has been reduced. The production of MBM from the specific risk material which has to be incinerated and from the dead animals takes place in one rendering plant, in which it is prohibited to render animal waste for feed production. The MBM from this plant is used in the cement industry as fuel.

For increased control, since July 1998 the organs to be incinerated must be dyed (8).

Although there has been no indication of a risk in tallow, a precautionary measure was introduced in September 1998; for feeding purposes, tallow has to be treated for 20 minutes at a temperature of 133°C (8).

Measures concerning human health

According to investigations in the UK, the possibility of vertical transmission could not be excluded (7, 39). Although in Switzerland no such indication was found, it was decided in September 1996 to dispose of all direct offspring of cows affected by BSE. Before this time, direct offspring of infected cows had to be tattooed and were banned for export.

Since January 1998, the production of mechanically recovered meat and gelatine from the head and vertebral column of cattle, sheep and goats has been prohibited.

Based on new scientific results, which indicated that infectivity may be found in dorsal root ganglia (25), the use of the bones of the vertebral column, the sacrum and the tail of cows as food was banned in July 1998; after heating at a temperature of 133°C and a pressure of 3 atmospheres for a minimum of 20 minutes, these materials may be used for animal feed, except for ruminants.

Import measures

As of May 1996, only imports of MBM without brain, spinal cord, eyes and dead animals, treated at a temperature of 133°C and a pressure of 3 atmospheres for a minimum of 20 minutes are allowed.

Since December 1996, the import of bovines, ovines and caprines is only permitted from countries which have introduced and effectively enforced a feed ban. As a rule, the animals must have been born at least 18 months after the feed ban. The import of meat products is only allowed from countries which do not use specific risk material.

Media and public reaction

The first four BSE cases in Switzerland were documented by press releases from the Swiss Federal Veterinary Office. Every further case was announced in the Bulletin, a communication paper which is published every fortnight by the Swiss Federal Veterinary Office. The first announcements resulted in a fairly intensive discussion in the media. However, the immediate measures enforced to limit the impact of BSE and the open information strategy of the Swiss Federal Veterinary Office calmed the reaction of the media and the public relatively quickly.

Until March 1996, the media in Switzerland reported sporadically on new cases in the country. However, the media response to the press releases and information from the Swiss Federal Veterinary Office was reserved.

The declaration of the authorities in the UK about the link between BSE and V-CJD on 26 March 1996 had a bombshell effect. The European beef market was in a dramatic situation. The export ban on British beef, imposed by the European Union, strengthened the anxiety of the consumer. Although the Swiss Federal Veterinary Office and the Swiss Public Health Office declared beef to be safe, Switzerland also implemented a complete ban on British beef as a political and confidence-building measure for the consumer.

In response to the crisis, the Swiss Federal Veterinary Office implemented a telephone 'hot-line'. The telephone lines rang all day for three weeks, then the situation calmed down.

The immediate reaction was a dramatic decrease in beef consumption. However, this was only a short-term reaction and the market rebounded rapidly (Fig. 5). A declining trend
in meat consumption in general has been seen over the years and the BSE scare did not make a conspicuous change to this trend (31).

Export trade impact
The reactions of importing countries were varied. After the announcement of the first case in Switzerland, only a few countries strengthened their import conditions in regard to Switzerland. However, after the declaration of a possible relation between BSE and V-CJD, many more importing countries reacted with import bans and more rigorous conditions. Most of the countries which did take measures did not base their import bans on the recommendations of the OIE, and in many cases the obligations of the World Trade Organization Agreement on Sanitary and Phytosanitary Measures were disregarded. While in some cases, ordinances or decrees were issued to provide a legal basis for the import ban, the veterinary administrations of other countries simply refused to issue import permits. Some countries restricted the import bans to live cattle, while others reacted with total import bans for any goods of bovine origin, including milk and semen, despite recommendations to the contrary (30). Some countries even prohibited transit of bovines through their territory.

Economic implications
The total expenses incurred by BSE in Switzerland are difficult to calculate but are surely not of the same magnitude as the losses in the UK. However, the economic consequences of BSE in Switzerland have been considerable. The costs of the disposal of BSE-affected animals and the costs of diagnosis have been paid by the cantons. The costs of culling the BSE herds, which accounted to more than SFr10 million (US$12.5 million), and of research and communication, which totalled approximately SFr4 million (US$5 million) were paid by the Swiss confederation. In addition to this, the time of those involved and export losses are difficult to estimate and have not been included here.

Conclusions
It seems that the number of cases of BSE is declining and the measures taken are effective. However, the potential weaknesses of current activities that allow the disease to exist are currently being evaluated. Further measures to avoid even minimal risk for human and animal health will be taken if necessary. Additional action for more intensive surveillance is currently being implemented; a representative sample of clinically healthy slaughter cows and dead cows are examined for subclinical BSE. Experience has shown that it is necessary to review the situation constantly and to inform the public and professionals regularly and in an open manner.

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L’encéphalopathie spongiforme bovine en Suisse : rappel historique et situation actuelle

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Résumé
Les mesures prises sont efficaces. Toutefois, au vu de la persistance de la maladie, les faiblesses potentielles des actions actuelles sont en cours d’évaluation. De nouvelles mesures visant à éviter tout risque, fût-il minime, pour la santé publique comme pour la santé animale seront prises si nécessaire. L’expérience montre qu’il convient de faire un bilan permanent de la situation et d’informer régulièrement le public et les éleveurs, dans un souci de transparence.

Mots-clés

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Resumen
En noviembre de 1990 se diagnosticó el primer caso en Suiza de encefalopatía espongiforme bovina, pese al hecho de que en dicho país no estuvieran presentes todos los factores de riesgo que fueron identificados durante la epidemia que afectó al Reino Unido. A 29 de diciembre de 1998 se habían registrado un total de 282 casos de la enfermedad (todos ellos en animales nacidos en Suiza). El número de casos sigue una tendencia descendente, con catorce casos diagnosticados durante 1998 (a fecha de 29 de diciembre).

Las medidas adoptadas hasta hoy resultan eficaces. Sin embargo, se están evaluando posibles puntos débiles de ciertas actividades que podrían facilitar la permanencia de la enfermedad. De ser necesario, se adoptarán nuevas medidas con objeto de evitar el menor riesgo para la salud humana y animal. La experiencia del pasado enseña que es necesario vigilar de modo permanente la situación y mantener informados al público y los profesionales de forma periódica y transparente.

Palabras clave
References


