

# SURVEILLANCE OF INFECTIOUS DISEASES

IN ANIMALS AND HUMANS IN SWEDEN 2022

*Chapter excerpt:  
Bovine spongiform encephalopathy*



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**Cover:** A cultivation of *Salmonella* at the Public Health Agency of Sweden.  
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**Reporting guidelines:** Reporting guidelines were introduced in 2018 for those chapters related to purely animal pathogens. The guidelines build on experiences from several EU projects, and have been validated by a team of international experts in animal health surveillance. The aim is to develop these guidelines further in collaboration within the global surveillance community and they have therefore been made available in the form of a wiki on the collaborative platform GitHub (<https://github.com/SVA-SE/AHSURED/wiki>). Feel free to contribute!

**Layout:** The production of this report continues to be accomplished using a primarily open-source toolset. The method allows the source text to be edited independently of the template for the layout which can be modified and reused for future reports. Specifically, the chapter texts, tables and captions are authored in Microsoft Word and then converted to the LaTeX typesetting language using a custom package written in the R software for statistical computing. The package uses the pandoc document conversion software with a filter written in the lua language. Most figures and maps are produced using R and the LaTeX library pgfplots. Development for 2022 has focused on generalising the R package to accommodate conversion into formats other than LaTeX and PDF, with a focus on markdown files which can be published as HTML websites using the Quarto publishing system. The report generation R package and process was designed by Thomas Rosendal, Wiktor Gustafsson and Stefan Widgren.

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# Bovine spongiform encephalopathy

## BACKGROUND

Classical bovine spongiform encephalopathy (BSE) belongs to a group of diseases called transmissible spongiform encephalopathies (TSEs). It was first described in cattle in the United Kingdom in 1986 and from there the disease spread to a large number of countries within as well as outside of Europe. The current theory about the causative agent is the protein-only hypothesis. This theory assumes that prions (misfolded and aggregated proteins) induce the same misfolded structure in normal prion-proteins in the body of the host, resulting in accumulation of prions and cellular damage without the involvement of any microorganism. Classical BSE primarily spread through animal feed containing contaminated meat and bone meal (MBM), i.e., MBM made from animals infected with BSE. However, the primary source of the epidemic was never established. The use of MBM was banned, first in feed to cattle and then in 2001, in feed to pigs and poultry to any avoid cross-contamination in the feed mills.

In 1996, the disease became a public health concern, following the detection of a new variant of Creutzfeldt-Jacob Disease in humans (vCJD) and a likely link to classical BSE in cattle. This resulted in actions taken to prevent transmission to humans, including the removal of specified risk material (such as brain and spinal cord) from cattle at slaughter

and feed restrictions to avoid possible recycling of infectious material to ruminants through infected MBM. Actions also included an intensified surveillance which started in 2001, when rapid diagnostic tests became available.

Atypical cases of BSE, which show diagnostic and epidemiological dissimilarities with classical BSE, were first described in the early 2000. These atypical BSE cases probably occur spontaneously (without known cause) and possible links to classical BSE and potential zoonotic aspects are subject to debate within the scientific community. Historically, the risk of BSE being introduced to Sweden has been considered low, as well as the risk of any recirculation of the disease, if it was to be introduced. This is due to an early ban on the use of fallen stock in the production of livestock feed, as well as limited imports. This is something that has been assessed by the Scientific Steering Committee, by the European Food Safety Authority (EFSA) (expressed in terms of the Geographical BSE Risk (GBR)) and later by the WOAHP Scientific Commission, Sweden is currently recognised as having a negligible risk for classical BSE, as a result of a resolution adopted by the WOAHP International Committee.

BSE has been detected in Sweden once, when a beef cow born in 1994 was found positive for atypical BSE of the H-type, i.e., not classical BSE, in 2006.



Figure 9: Classical bovine spongiform encephalopathy has never been detected in Swedish cattle. Photo: Bengt Ekberg/SVA.

## DISEASE

The incubation period of classical BSE is long, from two years up to several years. Clinical signs are related to the neurological system and include altered behaviour and sensation, as well as affected movement and posture. The clinical signs can last for weeks or months and the disease is progressive and always fatal.

## LEGISLATION

Surveillance and control of BSE is regulated through Regulation (EC) No 999/2001 of the European Parliament and of the Council of 22 May 2001. The surveillance design is in accordance with Annex III and Sweden applies derogation for remote areas with low cattle density (Commission Decision 2008/908) where there is no collection of fallen stock. The cattle population in these areas does not exceed 10% of the bovine population in Sweden. On the national level, the sampling is regulated by SJVFS 2010:9, last amended through SJVFS 2013:3. BSE is a notifiable disease under the Swedish Act of Epizootic diseases (SFS 1999:657, with amendments). Feed controls are regulated through Regulation (EC) 152/2009.

## SURVEILLANCE

### Feed

To investigate compliance with the feed bans, samples of feed and imported raw materials for feed production are collected at feed mills, points of retail and at farm level, and analysed for the presence of processed animal protein (PAP) using microscopy. This is part of the official control, and the Swedish Board of Agriculture as well as the County Administrative Boards are the responsible authorities. Samples are selected based on a risk assessment made by the Swedish Board of Agriculture.

### Animals

The Swedish Board of Agriculture oversee the surveillance programme which is carried out in cooperation with the National Veterinary Institute (SVA). Samples are analysed at SVA, which also is the National Reference Laboratory (Regulation (EC) 999/2001).

The risk of introduction and recirculation of BSE within the system has been controlled for many years. The purpose of the surveillance in animals is primarily to fulfil the requirements in the EU regulation and to show that Sweden can maintain the WOAHP status of negligible risk for classical BSE. WOAHP determines a minimum target for surveillance, which is based on a point system that needs to be reached for the preceding seven years. The points are allocated differently between different risk categories of animals, with so-called high-risk animals, such as animals with suspected clinical signs, rendering the highest number of points. The relative weight of different risk categories is based on historical BSE data from the United Kingdom.

### Passive surveillance

All clinical suspicions of BSE (bovine animals with clinical signs that are consistent with a BSE diagnosis, and not

responding to treatment) must be reported to the authorities. For the first part of 2022, samples from animals with a clinical suspicion of BSE were analysed with the TeSeE™ SAP rapid kit (Bio-Rad Laboratories, Hercules, California, United States). During 2022 the analysis method at SVA was changed, and for the second part of the year samples were analysed using the IDEXX HerdChek® BSE-Scrapie Antigen Test kit (IDEXX Laboratories, Westbrook, Maine, United States). In case of a positive or inconclusive result, the material is analysed with the TeSeE™ Western Blot kit (Bio-Rad Laboratories, Hercules, California, United States). Clinical suspicions are a category of animals that are important to include in the surveillance. However, since the control measures have been effective and the European epidemic of classical BSE has declined, it is a challenge to keep farmers and veterinarians alert and report these suspected clinical cases. Substantial efforts have been made, starting in 2018, to find animals that display clinical signs which could be compatible with BSE, and include these in the surveillance programme.

### Active surveillance

The following categories are sampled in the active surveillance (regulation 999/2001):

- Cattle of Swedish origin, above 48 months of age, that have remarks on antemortem inspection at slaughter, or are emergency slaughtered.
- Cattle of other than Swedish origin above 24 months of age that have remarks on antemortem inspection at slaughter or are emergency slaughtered.
- All healthy slaughtered cattle above 30 months of age that originate in a country other than Sweden, which does not have negligible risk for BSE.
- All fallen stock (animals dead or killed on farm but not slaughtered for human consumption) above 48 months of age that originate from Sweden. For cattle that originate from a country other than Sweden, which does not have a negligible risk for BSE, the age limit for sampling fallen stock is 24 months. Fallen stock are sampled by employees at the rendering plants, or by veterinarians or veterinary assistants at post-mortem examinations.

The diagnostic laboratory procedures are the same as for the passive surveillance (see above).

## RESULTS

### Feed

In 2022, 24 feed samples were taken at feed mills; 20 of these were from feed (17 were cattle feed) and four from raw material for feed production. All samples were negative for PAP, except two (one from pig feed and one from poultry feed), which contained fish particles.



## Animals

### *Passive surveillance*

In 2022, 8 bovines were examined due to clinical suspicion, all with negative results.

### *Active surveillance*

In 2022, 7654 samples were examined for BSE. All samples were negative. Out of these samples, 7420 were from fallen stock, 26 samples were from animals with remarks on ante-mortem inspection at slaughter and 200 samples were from emergency slaughtered animals.

## DISCUSSION

There were no cases of BSE detected in Sweden in 2022. The number of clinical suspicions has fluctuated over the years and has likely been related to the degree of awareness among farmers and veterinarians. A peak in suspected cases followed the BSE crisis, when media reporting on this disease was high. After a period of very few suspected clinical cases, there is again an increase as substantial efforts have been made to inform veterinarians of the importance of notifying the authorities of animals with clinical signs that could be compatible with BSE.

The source of the large epidemic of classical BSE has never been determined, and atypical cases cannot be excluded as the source. Atypical BSE cases could hence be a potential source of a new epidemic. As the number of cases of classical BSE is decreasing within the EU, surveillance is also declining. Suggestions have been made to again allow the use of MBM as well as processed animal protein (PAP) in feed within the EU. However, strict separation of feed materials during production, to avoid any cross-contamination, and bans of these feeding practices must be kept in place. This to avoid any possibility of BSE-prions recirculating if the disease agent was to again enter the feed system. It is essential that sampling of feed is done at sufficient levels to ensure compliance with the bans. However, the current number of samples is low, and the ability to detect a possible contamination in the feed system is therefore limited. Recent international reports of a few cases of classical BSE in young animals, born long after the implementation of the strict feed ban, either indicate problems with the ban, or

there are other causes of classical BSE that we do not yet understand.

A revision of the current surveillance requirements on EU and WOA levels is ongoing and motivated as the preventive measures that have been in place have resulted in a significant decrease of cases of classical BSE on a European and global level. It has been suggested to decrease the number of animals sampled, as large-scale sampling is not an efficient way to prevent a new BSE crisis. However, keeping feed bans and feed controls in place to avoid potential recirculation of prions is still relevant to avoid a new BSE epidemic.

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