

# SURVEILLANCE OF INFECTIOUS DISEASES IN ANIMALS AND HUMANS IN SWEDEN 2019

Chapter excerpt -  
Bovine spongiform encephalopathy



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**Reporting guidelines:** Reporting guidelines were introduced in 2018 for those 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, produced by authors, 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 using pandoc and R to the LaTeX typesetting language. Most figures and maps are produced using the R software for statistical computing. Development for 2019 has further improved the importing of content from Word to LaTeX. The method can now import text, tables and figure captions from Word, as well as the newly designed 'IN FOCUS' sections of some chapters. The tool is available as an R-package at GitHub (<https://github.com/SVA-SE/mill/>). This year the report was also built with a continuous integration pipeline on Microsoft's Azure DevOps platform, allowing every committed change to the content to be built and tested automatically. The report generation R-package and process was designed by Thomas Rosendal and Stefan Widgren. In 2019, figures and the final typesetting were done by Wiktor Gustafsson and Thomas Rosendal with contributions from the report authors.

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# Bovine spongiform encephalopathy

## BACKGROUND

Classical bovine spongiform encephalopathy (BSE) belongs to a group of diseases called transmissible spongiform encephalopathies (TSE). It was first described in cattle in the UK in 1986 and from there the disease spread to a large number of European countries as well as countries outside Europe. The current theory about the causative agent is the protein-only hypothesis. This theory assumes that misfolded prions (small 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 involvement of any microorganism. Classical BSE primarily spread through animal feed including contaminated meat and bone meal (MBM), i.e. MBM containing parts of animals infected with BSE. However, the primary source of the epidemic was never established. The use of MBM was prohibited, first in feed to cattle and in 2001 in feed to pigs and poultry to avoid cross-contamination in the feed mills.

In 1996, the disease became a public health concern, after the detection of a new variant of Creutzfeldt-Jacob Disease in humans (vCJD), likely to be linked to classical BSE in cattle. This resulted in actions taken to prevent transmission to humans through removal of specified risk material (such as brain and spinal cord) from cattle at slaughter, restrictions related to feed to avoid recycling of infectious material to ruminants through infected MBM and an intensified surveillance which started in 2001 after rapid diagnostic tests became available.

Atypical strains of BSE, which show diagnostic and epidemiological dissimilarities with classical BSE, have been described. These atypical BSE cases probably occur spontaneously and possible links to classical BSE and potential zoonotic aspects are subject to debate within the scientific community.

Sweden has historically had a low risk of introduction of classical BSE and a low risk of recirculation of the disease if it had been introduced, due to an early ban on the use of fallen stock in production of feed for livestock and limited imports. This 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 OIE Scientific Commission. Sweden is currently recognised as having a negligible risk for classical BSE, as a result of a resolution adopted by the OIE International Committee.

One case of BSE has been detected in cattle in Sweden. This was in 2006 in a beef cow born in 1994. This case was confirmed to be atypical BSE of the H-type, i.e. not classical BSE.

## DISEASE

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

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

### Animals

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

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 maintain the OIE status of negligible risk for classical BSE. The OIE 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 high risk animals, such as clinical suspicions, rendering the highest number of points. The relative weight of different categories is based on historical BSE data from the United Kingdom.

### Passive surveillance

All suspicions of BSE (bovine animals not responding to treatment, with clinical signs that are consistent with a BSE diagnosis) must be reported to the authorities. The obligation to report applies to animal owners, veterinarians and everyone else who is responsible for the animals. If the animal is still alive, it is examined by a veterinarian who is in close contact with disease experts and it is decided if the animal should be euthanized. Samples are analysed with Bio-Rad TeSeE short assay protocol (SAP). In case of positive or inconclusive results, the material is prepared and examined



with Bio-Rad TeSeE Western Blot.

Clinical suspects are a category of animals that should be included in the surveillance, but 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 symptoms. Substantial efforts have been made starting 2018 and continuing 2019 to find animals that display symptoms which could be compatible with BSE and to include these in the surveillance programme.

#### *Active surveillance*

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

- Cattle of Swedish origin, above 48 months of age, that have remarks at antemortem inspection before slaughter or are emergency slaughtered.
- Cattle of other than Swedish origin above 24 months of age that have remarks at antemortem inspection before 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. The fallen stock are sampled by employees at the rendering plants or by veterinarians or veterinary assistants at postmortem.

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

## RESULTS

### **Feed**

In 2019, 22 feed samples were taken at feed mills; 19 of these were from feed (13 were cattle feed) and three from raw materials for feed production. All of these samples were negative for PAP, except one feed which contained fish particles, but it was a feed for pigs which included fish meal in the recipe.

### **Animals**

#### *Passive surveillance*

In 2019, 18 bovines were examined due to clinical suspicion, all with negative results.

#### *Active surveillance*

In 2019, 8423 samples were examined for BSE. All samples were negative. Of these samples 8226 were from fallen stock, 26 samples were from animals with remarks at antemortem inspection before slaughter and 152 samples were from emergency slaughtered animals.

## DISCUSSION

No positive BSE cases were detected in Sweden in 2019. Preventive measures have been in place for many years and

the fact that no cases were detected supports that these measures have been effective. The increased number of clinical suspicions in 2018 and 2019 compared to previous years is the result of substantial efforts to detect and notify animals with clinical signs compatible with BSE. There has previously been a long trend of decreasing number of clinical suspicions compared to the years during the peaks of the BSE crisis, which can be explained by to a lower degree of awareness among farmers and veterinarians when there is less reporting about the disease.

Reports of prion transmission studies, including several passages in different species, have shown that prion strains do not always remain stable through these passages. The source of the large epidemic of classical BSE has not been determined and atypical cases cannot be excluded as the source. Thus, the atypical cases could be a potential source of a new epidemic. As the number of cases of classical BSE is decreasing within the European Union, surveillance is decreasing, and suggestions have been made to once again allow the use of MBM in feed within the EU. However, strict separation and bans of these feeding practices must be kept in place to avoid any possibility of recirculation of BSE, if the disease agent were to enter the system again. Sampling of feed needs to be at sufficient levels to ensure compliance with bans; SVA would welcome increased sampling in the feed chain in Sweden. Recent international reports of a few cases of classical BSE in young animals, born long after implementation of the strict feed ban, either indicates problems with the ban, or there are other causes of classical BSE that we do not yet understand.

On OIE and European union level, a revision of the current surveillance of animals is being discussed, and a revision is welcomed. Spending large resources on sampling animals is not the most efficient way to prevent a new BSE crisis. But keeping bans and controls in place to avoid recirculation is still relevant to avoid a new BSE crisis.

## REFERENCES

Gavier-Widén D, Nöremark M, Langeveld JP, Stack M, Biacabe AG, Vulin J, Chaplin M, Richt JA, Jacobs J, Acín C, Monleón E, Renström L, Klingeborn B, Baron TG (2008) Bovine spongiform encephalopathy in Sweden: an H-type variant., *J Vet Diagn Invest* 20:2–10.

Capobianco R, Casalone C, Suardi S, Mangieri M, Miccolo C, Limido L, Catania M, Rossi G, Di Fede G, Giaccone G, Bruzzone MG, Minati L, Corona C, Acutis P, Gelmetti D, Lombardi G, Groschup MH, Buschmann A, Zanusso G, Monaco S, Caramelli M, Tagliavini F (2007) Conversion of the BASE prion strain into the BSE strain: the origin of BSE? *PLoS Pathog* 3(3):e31.

EFSA Panel on Biological Hazards, 2004. Scientific Report of the European Food Safety Authority on the Assessment of the Geographical BSE Risk (GBR) of Sweden. *EFSA Journal* 2004; 2(8):RN-7, 27 pp. <https://doi.org/10.2903/j.efsa.2004.7r>