

# SURVEILLANCE OF INFECTIOUS DISEASES

IN ANIMALS AND HUMANS IN SWEDEN 2022

*Chapter excerpt:  
Chronic wasting disease*



**Editor:** Karl Ståhl

Department of Epidemiology and Disease Control  
National Veterinary Institute (SVA), SE-751 89 Uppsala, Sweden

**Authors:** Emmi Andersson, Märit Andersson, Charlotte Axén, Anna Bonnevie, Ioana Bujila, Erika Chenais, Mariann Dahlquist, Leigh Davidsson, Rikard Dryselius, Helena Eriksson, Linda Ernholm, Charlotta Fasth, Malin Grant, Gittan Gröndahl, Gunilla Hallgren, Anette Hansen, Marika Hjertqvist, Mia Holmberg, Cecilia Hultén, Hampus Hällbom, Helena Höök, Karoline Jakobsson, Désirée Jansson, Tomas Jinnerot, Jonas Johansson Wensman, Jerker Jonsson, Oskar Karlsson Lindsjö, Sara Kjellsdotter, Ulrika König, Elina Lahti, Emelie Larsdotter, Neus Latorre-Margalef, Mats Lindblad, Anna Lundén, Anna Nilsson, Oskar Nilsson, Maria Nöremark, Anna Omazic, Anna Ordell, Ylva Persson, Emelie Pettersson, Ivana Rodriguez Ewerlöf, Thomas Rosendal, Marie Sjölund, Karl Ståhl, Lena Sundqvist, Robert Söderlund, Magnus Thelander, Karin Troell, Henrik Uhlhorn, Anders Wallensten, Stefan Widgren, Camilla Wikström, Ulrika Windahl, Beth Young, Nabil Yousef, Siamak Zohari, Erik Ågren, Estelle Ågren

**Typesetting:** Wiktor Gustafsson

**Cover:** A cultivation of *Salmonella* at the Public Health Agency of Sweden.  
Photo: Nicklas Thegerström/DN/TT. Cover design by Rodrigo Ferrada Stoeהל.

**Copyright of map data:** ©EuroGeographics for the administrative boundaries

**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.

**Print:** TMG Tabergs AB

Except where otherwise noted, the reuse of this document is authorised under the Creative Commons Attribution 4.0 International (CC BY 4.0) licence. This means that reuse is allowed provided appropriate credit is given and any changes are indicated. For any use or reproduction of photos or other material that is not owned by SVA, permission must be sought directly from the copyright holders.

**Suggestion citation:** Surveillance of infectious diseases in animals and humans in Sweden 2022, National Veterinary Institute (SVA), Uppsala, Sweden. SVA:s rapportserie 89 1654-7098

This report may be subject to updates and corrections. The latest version is always available for download at [www.sva.se](http://www.sva.se).



# Chronic wasting disease

## BACKGROUND

Chronic wasting disease (CWD) is a transmissible spongiform encephalopathy (TSE) affecting cervid species. The disease was first described in Colorado in 1967 and in 1978 it was identified as a TSE. Since then, CWD has spread and is now confirmed to be present in at least 30 states in the USA, and in four Canadian provinces (USGS, 2022). Through export of live cervids, CWD has also spread to South Korea.

Prior to 2016, CWD had not been reported in Europe but in the spring of 2016, the first case in Europe was detected in wild reindeer (*Rangifer tarandus tarandus*) in the region of Nordfjella in Norway (Benestad et al., 2016). As a consequence of this finding, surveillance in Norway was intensified and this has so far (April 2023) resulted in the detection of the disease in 21 reindeer in two different wild reindeer areas. In addition, CWD has been found in 11 elderly moose (*Alces alces*) and in three red deer (*Cervus elaphus*) in different locations. The reindeer in Norway show similarities with the cases found in North America with several animals affected in the same flock, and with lymph nodes being positive on analysis. The cases in moose and red deer have however shown to differ from the cases in reindeer. In moose

and red deer, the positive cases have been found in older animals and samples have tested positive from the brain, but not from the lymph nodes. Following the detection of CWD in Norway, surveillance for CWD became mandatory in several EU member states, including Sweden in the time period 2018–2020 (see sections “Legislation” and “Surveillance”).

In March 2018, the first case of CWD in Finland was reported in a moose. The case showed similarities with the cases in moose and red deer in Norway (Ruokavirasto, 2020). Two similar cases were reported from Finland in 2020 and in 2022.

In Sweden the first three cases were detected in 2019 and all were elderly female moose. The first two animals (one found in Arjeplog and one in Arvidsjaur) were both euthanised and sampled after displaying abnormal behaviour. Both were estimated to be 16 years old. In accordance with EU legislation, an intensified sampling was carried out in the area during the following hunting (moose) and slaughter (reindeer) seasons and the third case, a female of at least 10 years of age, was detected within this intensified sampling. Although shot during the regular hunting, the moose was showing altered behaviour and standing still in a mire.

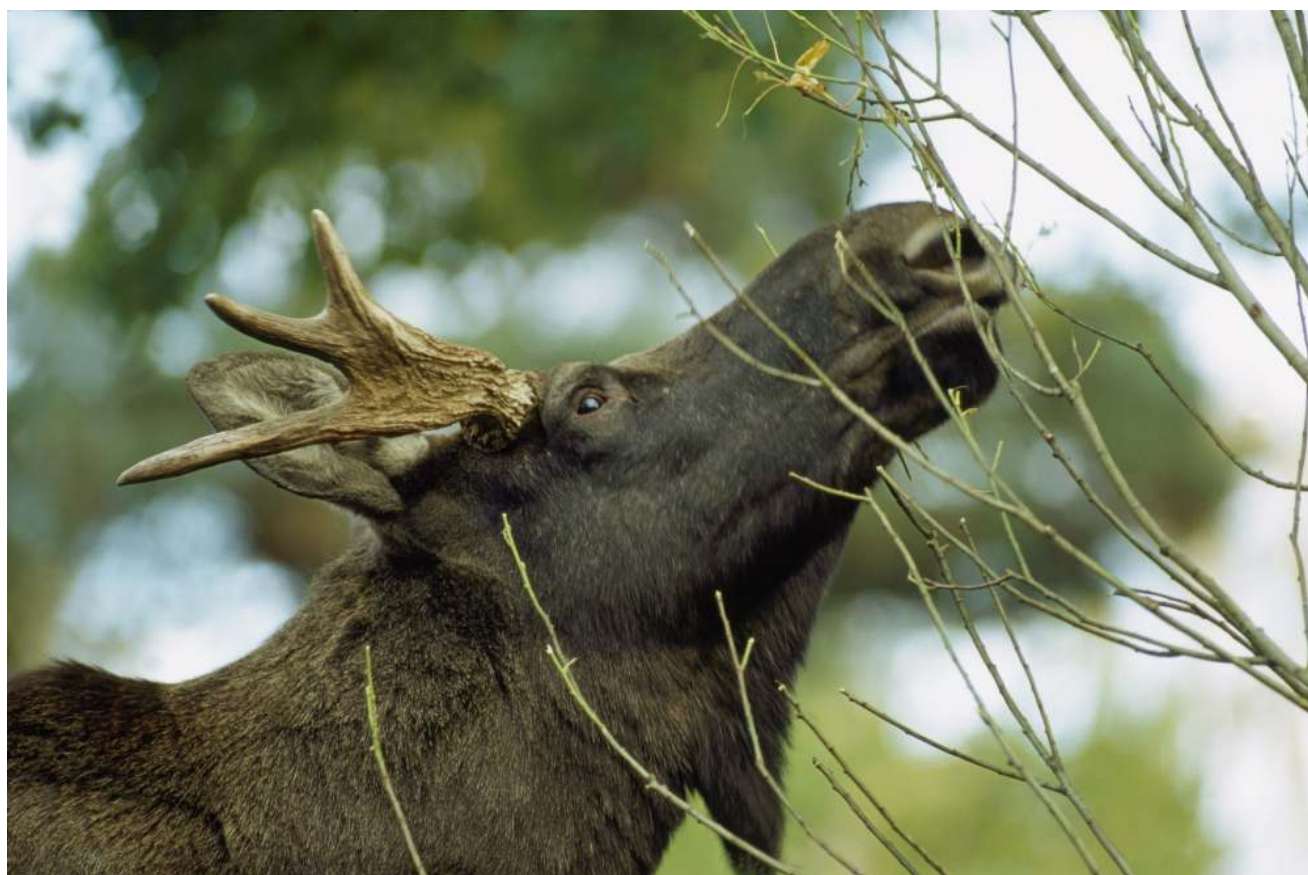


Figure 15: The EU-regulated surveillance of chronic wasting disease (CWD) was discontinued in March 2022, but the passive clinical surveillance continues. Photo: SVA.

A fourth case was detected in 2020 in the Robertsfors municipality. It was in a female moose aged to 14 years (age estimated by counting annuli in dental cementum), shot as she had been observed with lameness and when approached by hunters, she did not attempt to escape. On analysis, samples were positive from the brainstem but negative from the lymph nodes.

These four cases all showed similar features with the cases that had been reported from moose in Norway and in Finland. All were in old moose, all positive in the brainstem sample but negative in the lymph-node samples. These findings in moose differ significantly from the findings in cervids in North America and wild reindeer in Norway and it has been hypothesised that these cases may be occurring spontaneously in older animals (Pirisinu et al., 2018).

The wild cervid species in Sweden are moose, red deer, fallow deer (*Dama dama*) and roe deer (*Capreolus capreolus*), and hunting of these species involve and engage many people. The farmed cervid species in Sweden are mainly fallow deer and smaller number of red deer and moose. In Sweden, reindeer herding is an essential part of the Sami culture, all reindeer are semi-domesticated and unlike in Norway, there are no wild reindeer. However, the cervid populations are not separated by geographical borders, and both wild cervids and semi-domesticated reindeer cross the border between Sweden and Norway as part of for example the seasonal migration.

## DISEASE

From what is known about the strains of CWD present in North America, the incubation period is long, i.e., more than one year. The disease spreads either through direct contact between animals, or through excretions that can contaminate and persist in the environment. The predominant clinical signs are behavioural changes, change of locomotion and loss of body condition. The disease is always fatal.

Until recent years, the type of CWD described from North America was the only type known. The strains detected in the Nordic countries, however, differ from the strains described from North America. There is accumulating support for the hypothesis that the cases seen in older moose may have a spontaneous (i.e., without known cause), rather than contagious origin, similar to what is observed in sheep (atypical scrapie/Nor98) and bovines (atypical bovine spongiform encephalopathy (BSE)), and sporadic Creutzfeldt-Jakob disease (CJD) in humans.

The currently accepted theory of TSEs, or prion diseases, is that they are transmitted through small prions (aggregated proteins with abnormal structural conformation). These prions induce a structural transformation and aggregation of normal prion proteins in the body of the recipient. The full details of these processes are not yet understood. Prions accumulate in body tissues, especially in the brain where damage can be observed if tissue is studied in a microscope. Within the TSE group of diseases, there are diseases where prions are excreted in body fluids (e.g., classical scrapie) and hence these are contagious. However, there are also prion diseases with spontaneous (without known cause)

origin occurring in older individuals (e.g., atypical BSE in bovines or sporadic CJD in humans).

Due to similarities with BSE, a disease which is linked to variant CJD in humans, and the known fact that many TSEs experimentally can be transmitted between different species, there has been a suspicion that CWD may be zoonotic. Currently, there is not enough data to exclude that CWD could be zoonotic, however the risk is deemed to be very low (VKM 2016 and 2017, Waddell 2018). In areas where CWD is endemic, people are recommended to not consume animals displaying clinical signs consistent with CWD, or animals that have tested positive for CWD.

## LEGISLATION

CWD is a notifiable disease under the Swedish Act of Epizootic diseases (SFS 1999:657, with amendments) and there is a scheme to compensate farmers and owners for losses due to eradication measures. CWD is also regulated through the Regulation (EC) No 999/2001 of the European Parliament and of the Council on TSEs.

## SURVEILLANCE

### Passive surveillance

Since 2016, efforts have been made to increase awareness among hunters, reindeer owners and owners of farmed deer, to react and report suspected clinical signs of CWD. This has still been encouraged in 2022, when the active surveillance programme was discontinued. In addition to clinical suspicions, adult cervids sent for autopsy to the National Veterinary Institute (SVA) are sampled as part of an enhanced passive surveillance.

### Active surveillance

The time-limited EU-regulated surveillance programme started in January 2018. For the member states concerned, a minimum of 6000 animals were to be tested between the years 2018 and 2020. Samples were collected from wild, semi-domesticated and farmed/captive cervids. In Sweden, moose, red deer, roe deer and reindeer were included in the surveillance programme. Samples from wild cervids (moose, red deer, roe deer) were collected from fifty primary sampling units (PSU) covering the whole country. Samples from farmed cervids (red deer) and semi-domesticated cervids (reindeer) were collected from all red deer farms (n=137) and all Samebyar (n=51), the latter being the administrative unit for reindeer herding, as well as a geographically defined area. All animals sampled were over twelve months of age and preferably from a risk category, i.e., cervids found dead or diseased, or road/train killed cervids which are assumed to have a higher probability of infection. Because the sampling target had not been reached by the end of 2020, the surveillance was extended throughout 2021 and early 2022. In March 2022 the active surveillance programme was discontinued.

### Intensified active surveillance

In accordance with EU legislation, an intensified sampling was carried out in the areas surrounding the previously confirmed cases. The size of the areas was determined by experts in cervid migratory patterns and experts in reindeer herding. The surveillance was designed to enable detection of a prevalence of 0.7% (detected prevalence in Nordfjella, Norway) with 99% certainty, assuming a test sensitivity of 70%, in each population in the area. In this sampling, healthy slaughtered and healthy hunted animals were also included to increase the number of tested animals in the area. In 2019–2021 (moose 2019–2020, reindeer 2019–2021) intensified sampling was done around the confirmed cases in Arvidsjaur and Arjeplog, and in 2020–2021 Robertsfors was also included.

### Diagnostics

All samples were analysed at SVA, Uppsala, which also is the national reference laboratory (Regulation (EC) 999/2001) for TSEs. For analysis, brainstem and retropharyngeal lymph node samples were for the first part of 2022 screened separately with the Rad TeSeE™ SAP rapid test (Bio-Rad Laboratories, Hercules, California, United States). During 2022 the analysis method at SVA was changed, and the HerdChek® BSE-Scrapie Antigen Test kit (IDEXX Laboratories, Westbrook, Maine, United States) was used. If results are positive or inconclusive the Bio-Rad TeSeE™ Western Blot kit is used for confirmation. Test results are reported to the European Food Safety Authority by the Swedish Board of Agriculture (SBA), based on data provided by SVA.

The 2018–2022 CWD surveillance programme was run in a collaboration between SVA and the SBA and was financed by the latter.

## RESULTS

The number of samples tested from 2016 to 2022 is detailed in Table 7.

In 2022, 19 cervids were sampled due to a reported clinical suspicion of CWD (16 moose and 3 roe deer). An additional 12 moose were examined within the framework for enhanced passive surveillance. In the active surveillance programme, a total of 12 moose, 100 red deer, 1 roe deer and 502 reindeer were examined for CWD at SVA during 2022. No positive cases were found during 2022.

## DISCUSSION

With the exception of an EU regulated active surveillance in 2007–2010, and a retrospective study examining 270 frozen brains from cervids sent for post-mortem investigation between 2008 and the first part of 2016, surveillance in Sweden has historically only been passive, i.e., based on reporting of animals showing clinical signs. However, since the disease was a not known to occur in Europe prior to 2016, the awareness of CWD was low and consequently very few animals have historically been examined.

Before 2018, the number of animals examined was

limited and not well represented geographically. In January 2018, the surveillance programme (Regulation (EC) 999/2001) started and information about this was sent out to groups identified as suitable samplers, i.e., hunters, animal owners (reindeer and fenced red deer) and slaughterhouse personnel.

The number of samples received from the nationwide sampling has however been relatively low and there are several reasons for this. The implementation of the surveillance programme has been complex, given several different species and categories of animals included. Also, the number of animals found dead or diseased, which are preferred due to their assumed higher probability of infection, has been relatively small. The number of road-killed cervids is quite high in Sweden and they were initially planned to constitute the backbone of the surveillance. However, divergent interpretations of legislation made sampling of these animals difficult or even impossible. As the sampling targets were not reached the national surveillance was prolonged until March 2022, with the aim to collect more samples. In contrast, sampling of healthy hunted moose and healthy slaughtered reindeer within the intensified surveillance has been more successful despite logistical challenges.

As mentioned, the cases in moose in Norway, Finland and Sweden differ from the CWD cases previously known; all have been detected in old female animals and prions have only been detected in samples from the brain and not in lymph nodes. Further studies are ongoing to characterise these strains and to understand more about the epidemiology. As mentioned, it has been hypothesised that the CWD cases in older moose may not be contagious, but rather a spontaneous variant of CWD occurring in old animals (Pirinu et al., 2018).

Consequently, the detection of three cases of CWD in moose in a limited geographical area in Sweden in 2019 (detected prevalence 0.4%) does not necessarily mean that a contagious variant of the disease is present in the region. The fact that cases were only found in old animals and that prions were only detected in brain in the screening tests, while not in lymph nodes, still fits the hypothesis of spontaneous (without known cause) cases in old animals. The region of concern has a hunting practice which leads to a relatively high proportion of old female moose in the local population. In general, most moose are harvested at a young age during hunting, and few animals reach the expected maximum life span of approximately twenty years. Fewer males reach an old age compared to females due to the hunting practices.

The experience from North America is that CWD is very difficult to eradicate or to control, and to have a chance to do this, early detection is needed while the prevalence is still low. If a type of CWD with these characteristics would be present or introduced into Sweden, it would potentially have large negative consequences for reindeer, wild cervid populations and farmed cervids. Consequently, the disease could also have large consequences for people making their living from, or being involved in activities related to, these species.



Table 7: The number of animals tested for CWD per year in Sweden 2016–2022, including national surveillance and intensified sampling.

Year	Moose	Red deer	Roe deer	Fallow deer	Reindeer
2016	74	6	14	0	2
2017	191	6	13	8	21
2018	157	13	15	0	15
2019	854 <sup>A</sup>	31	73	5	1965 <sup>A</sup>
2020	248 <sup>B</sup>	84	71	4	991 <sup>C</sup>
2021	433 <sup>B</sup>	290	63	3	2527 <sup>C</sup>
2022	40	100	4	0	502 <sup>C</sup>

<sup>A</sup> The large increase in sampling in 2019 was due to the intensified sampling in the county of Norrbotten, which started that year.

<sup>B</sup> 98 and 283 of the moose sampled in 2020 and 2021, respectively, were sampled within the intensified sampling in the county of Västerbotten.

<sup>C</sup> 896, 1485 and 502 of the reindeer sampled in 2020, 2021 and 2022 respectively were sampled within the intensified sampling in the county of Norrbotten.

However, if the cases found in older moose in Norway, Finland and Sweden would in fact have a spontaneous (without known cause) origin, the disease could be expected to occur sporadically in all cervid populations, without leading to the same severe consequences as a contagious form of CWD. Further studies are crucial to increase the understanding of the epidemiology of the different CWD types.

## REFERENCES

- U.S. Geological Survey and Centers for Disease Control and Prevention, <https://www.cdc.gov/prions/cwd/occurrence.html> <https://www.usgs.gov/media/images/distribution-chronic-wasting-disease-north-america-0> (accessed 2023-04-19)
- Benestad SL, Mitchell G, Simmons M, Ytrehus B, Vikøren T (2016) First case of chronic wasting disease in Europe in a Norwegian free-ranging reindeer. *Vet Res* 47:88
- Nonno R, Di Bari MA, Pirisinu L, D'Agostino C, Vanni I, Chiappini B, Marcon S, Riccardi G, Tran L, Vikøren T, Våge J, Madslie K, Mitchell G, Telling GC, Benestad SL, Agrimi U. Studies in bank voles reveal strain differences between chronic wasting disease prions from Norway and North America. *Proc Natl Acad Sci U S A*. 2020 Dec 8;117(49)
- Pirisinu L, Tran L, Chiappini B, Vanni I, Di Bari MA, Vaccari G, Vikøren T, Madslie KI, Våge J, Spraker T, Mitchell G, Balachandran A, Baron T, Casalone C, Rolandsen CM, Røed KH, Agrimi U, Nonno R, Benestad SL (2018) Novel Type of Chronic Wasting Disease Detected in Moose (*Alces alces*), Norway. *Emerg Infect Dis* 24:2210–2218
- Ruokavirasto, <https://www.ruokavirasto.fi/viljelijat/elaintenpito/elainten-terveys-ja-elaintaudit/elaintaudit/luonnonvaraiset-elaimet/hirvielainten-naivetystauti-cwd/> (accessed 2020-03-16)
- VKM (2016) CWD in Norway. Opinion of the Panel on biological hazards, ISBN: 978-82-8259-216-1, Oslo, Norway
- VKM (2017) CWD – update statement. Opinion of the Panel on biological hazards, ISBN: 978-82-8259-283-3, Oslo, Norway
- Vikøren T, Våge J, Madslie KI, Røed KH, Rolandsen CM, Tran L, Hopp P, Veiberg V, Heum M, Moldal T, Neves CGD, Handeland K, Ytrehus B, Kolbjørnsen Ø, Wisløff H, Terland R, Saure B, Dessen KM, Svendsen SG, Nordvik BS, Benestad SL. First Detection of Chronic Wasting Disease in a Wild Red Deer (*Cervus elaphus*) in Europe. *J Wildl Dis*. 2019 Oct;55(4):970–972. Epub 2019 Mar 28. PMID: 30920905
- Waddell L, Greig J, Mascarenhas M, Otten A, Corrin T, Hierlihy K (2018) Current evidence on the transmissibility of chronic wasting disease prions to humans-A systematic review. *Transbound Emerg Dis* 65:37–49
- Ågren EO, Sörén K, Gavler-Widén D, Benestad SL, Tran L, Wall K, Averhed G, Doose N, Våge J, Nöremark M. First Detection of Chronic Wasting Disease in Moose (*Alces alces*) in Sweden. *J Wildl Dis*. 2021 Apr 1;57(2):461–463. doi: 10.7589/JWD-D-20-00141. PMID: 33822167