Epidemiology and animal TSEs





outline

- Animal TSEs: epidemiologically heterogeneous
- Classical BSE: the disappearance of a non-contagious transmissible disease
- Classical scrapie in small ruminants: successful genetic strategies to contrast transmissibility
- Chronic Wasting Disease in Cervids: controllable?
- Sporadic TSEs with unknown risk factors: diseases so rare as to prevent understanding?

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Prion disorders and epidemiology

- Familial diseases, i.e. genetic: Fatal Familial Insomnia
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classical BSE

BSE: Classical and atypical forms

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Atypical forms (L-BSE and H-BSE), identified by chance during surveillance (2004), are similar but affect much older cows, their causes are unknown and they are very, very rare

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The epidemiological cycle of BSE: This is why today we are here all together!



Transmissible but non contagious...

MBM import Cattle import



Cross-contamination: how?



in Hornlimann et al. 2006

The UK

Ireland

France



The Nethrlands

Germany

Ducrot et al., 2010



Worldwide, since the first 2000 decade the epidemics have been fading out!!!

This is the result of the application of evidence based effective measures to control the disease





Mad cow disease, a disease now trying to go unnoticed



Mad cow disease, a disease now trying to go unnoticed



Mad cow disease, a disease now trying to go unnoticed



The BARB issue



EFSA Panel on Biological Hazards (BIOHAZ),

Born After the Reinforcement of the Ban (BARB)

...contaminated feed is [still] the most likely source of infection

[...] the infectious agent [...] has the ability to remain active for many years.

[...] the BSE infectious agent was present where feed was stored or handled.

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Classical scrapie

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Effectiveness of national breeding plans





Time (caseload 2013-22)



Space (caseload in 2022)







Efsa, 2023



■ NSP1 ■ NSP2 ■ NSP3 ■ NSP30 ■ NSP4 ■ NSP5

	2013	2014	2015	2016	2017	2018	2019	2020	2021	2022
Number of contributing MS	24	25	25	25	20	7	7	8	8	7
Total genotyped	8,340	9,437	9,823	9,413	8,871	2,713	3,180	7,985	8,068	7,904



Genotyping in sheep: random (2012-2022)

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CWD

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		Reindeer	Moose	Red deer	
(https://www.efsa.europa.eu/en/efsajournal/pub/7936)	Age	Young/adult (mid age)	High	High	
Ly+ vs. Ly-	Distribution of prions	Lymph nodes alone or also brain	In brain but not in lymph nodes	In brain but not in lymph nodes	
2017-2022 monitoring	Distribution in brain	Disseminated, mostly brainstem	Mostly in neurones, whole brain	Intra, extraneuronal, star- like etc	
8 northern countries155,660 tests					
 83.8% HSHC ++ Road Kills 	Western blot	Indistinguishable from findings in USA	Lower MW, N-terminal truncated	Intermediate MW, N-terminal truncated,	
 ++ Roe Deer CWD in Sweden 			Rein Red D Mo BSE	BSE-like profile	
(moose) & Finland (moose)					
		Ly+	-	Ly-	

Geographical distribution by species in **Norway**

3

21





2019



CWD cases by species and country (2016-2024). Modified from EFSA <u>https://bit.ly/48u4wx3</u>

-p

REINDEER



Ly+





CWD cases by species and country (2016-2024). Modified from EFSA <u>https://bit.ly/48u4wx3</u>

NORWAY



Modified from EFSA <u>https://bit.ly/48u4wx3</u>



CWD cases by species and country (2016-2024). Modified from EFSA <u>https://bit.ly/48u4wx3</u>

NORWAY

Geographical distribution by species in Norway, **Sweeden and Finland**













CWD cases by species and country (2016-2024). Modified from EFSA <u>https://bit.ly/48u4wx3</u>

Geographical distribution by species in **Norway**

3

21



Live TV Opinion Diaspora India Global #RaceToWhiteHouse

News > World News > 'Zombie Deer Disease' Hits Another US State As 2 White-Tailed Deers Test Positive In Virginia

NDTV

'Zombie Deer Disease' Hits Another US State As 2 White-Tailed Deers Test Positive In Virginia

Los Angeles Times

CLIMATE & ENVIRONMENT



The age of extinction is supported by

the guardian

org

About th

Todd Bozel

Fri 22 D

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British Columbia 2 cases of 'zombie deer disease' have been confirmed in B.C. Here's what you need to know

• This article is more than **2 months old**

'Zombie deer disease' epidemic

spreads in Yellowstone as scientists

raise fears it may jump to humans

Hunters warned to not eat meat of any animals exhibiting symptoms CBC News - Posted: Feb 01, 2024 6:11 PM EST | Last Updated: February 10



Deadly brain disease found in two California deer **Breaking news!**









four years later...









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L & H BSE, scrapie NOR98, and Ly- CWD

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•2022 • H-BSE

• France

•2023

• H-BSE

- Brazil
- Spain
- the UK

• 2024

• H-BSE

- Brazil
- Spain

•L-BSE:

- the Netherlands
- Spain
- Switzerland
- US

- C-BSE:
 - the UK

Time (cases/million tests 2017-22)



Space (cases/million tests 2001-22)



Efsa, 2023

NOR98 (atypical scrapie) in small ruminants



Figure 2: Number of cases of AS reported by EU MS during the period 2003–2019 for sheep and 2005–2019 for goats

Time (caseload 2013-22)

Space (caseload in 2022)



Efsa, 2023

Prion disorders and epidemiology

- Sporadic diseases, unknown risk factors
- Few sources of data and those present are progressively disappearing...
- Diseases so rare as to prevent understanding their epidemiology and risk factors?
- Often they are considered **spontaneous** (=no risk factors)
- An easy answer? would this justify inaction?

Prion disorders and epidemiology



[...] this review discusses limitations of traditional approaches and advantages of molecular epidemiology approaches to investigate sporadic and endemic infections. Using specific examples, the discussions show that most sporadic infections are actually part of unrecognized outbreaks and that what appears to be endemic disease occurrence is actually comprised of multiple small outbreaks.