

CWD in Norway 2016; status and research needs



Deputy director Jorun Jarp

European Animal health and welfare research; collaboration research group

17th of November 2016, London

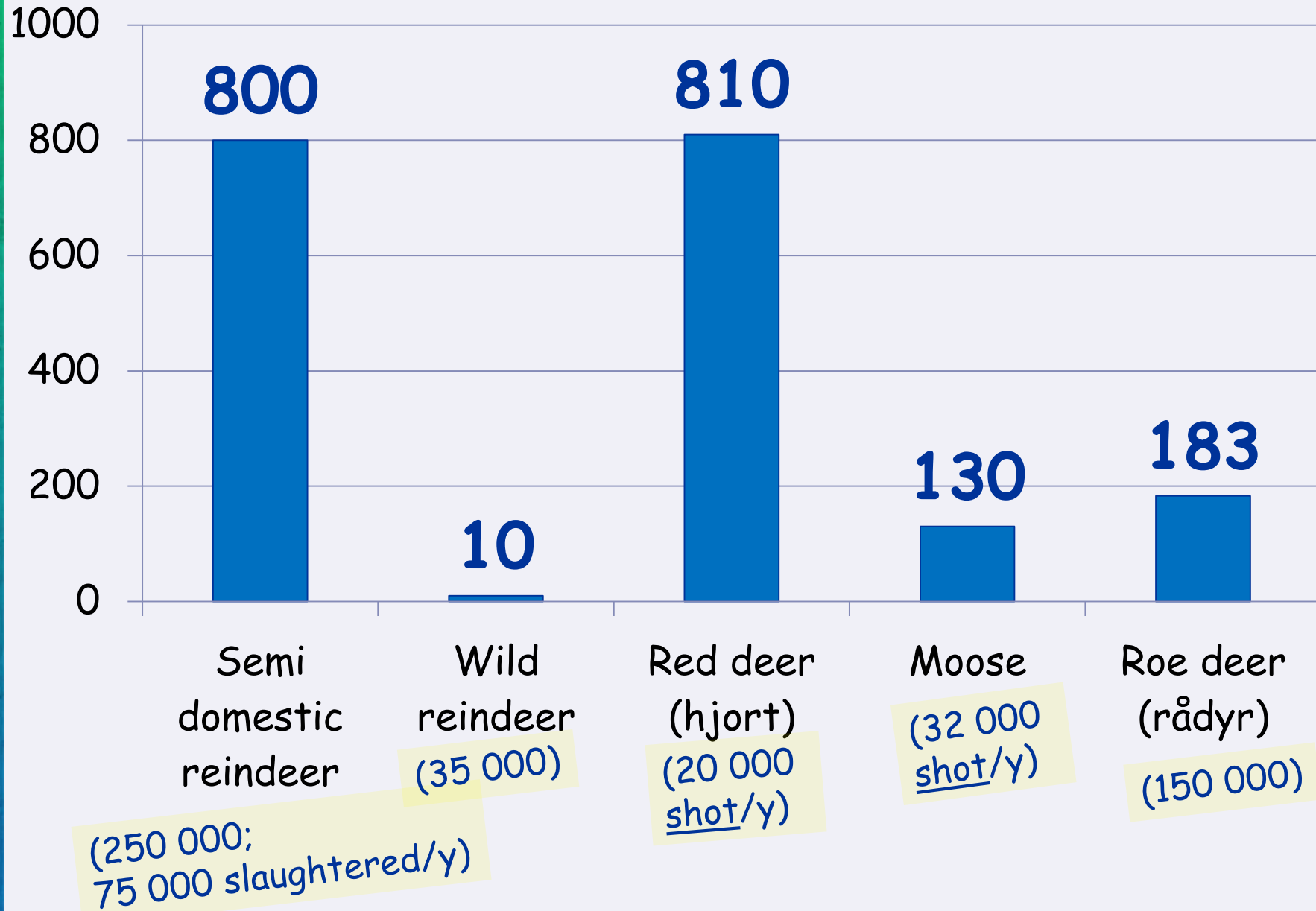
Cases of CWD documented in Norway so far

- Three wild reindeers from a stock in Nordfjella
- Two adjacent moose found in the middle of Norway





Analysis of cervids for CWD 2004-2015 per species- Total 1943 cervids



Wild reindeer, flock of 400, Nordfjella
GPS-collaring project- NINA
Helicopter
Isolated, tranquilised?



→ Locomotions problems?



Photo: NINA



illustration: The Gardian



Clinical status



ELISA
(brain)



Easter 2016: Test TeSeE ELISA
SAP Bio-Rad
High positive

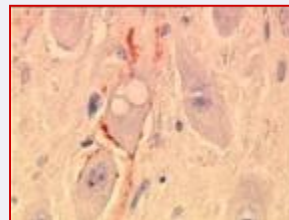
PrP^{Sc} western blot
(brain)



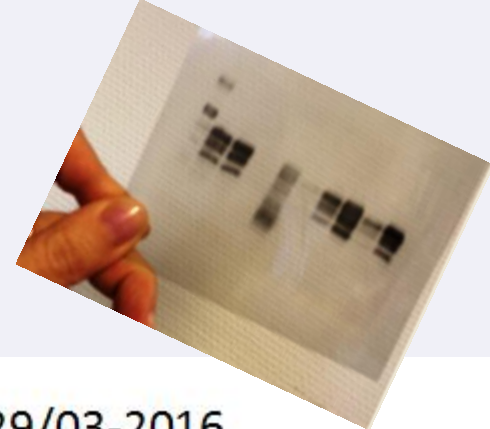
TSE
Diagnosis
Scrapie, BSE,
CWD

PrP^{Sc} immunohistochemistry
(brain)

research →

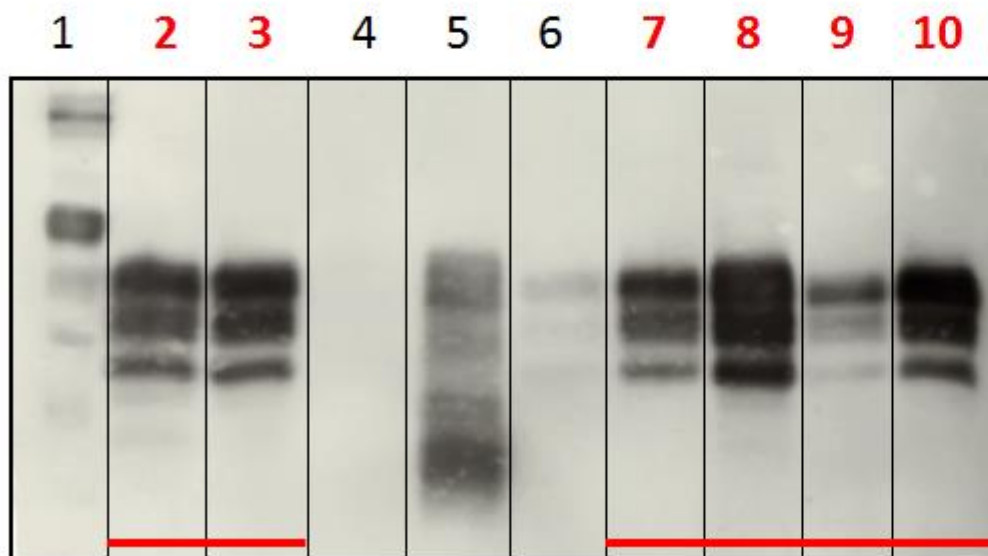


Confirmed positive with WB



WB 345

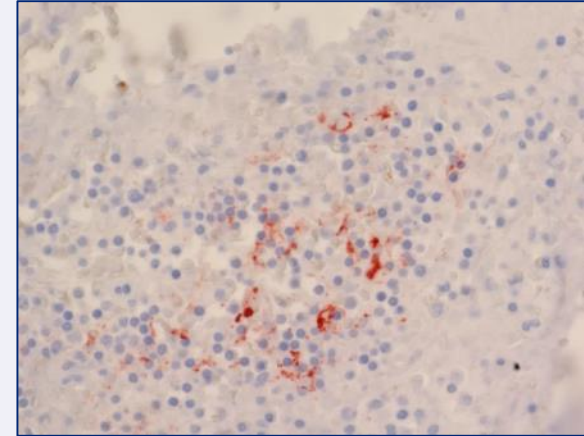
29/03-2016



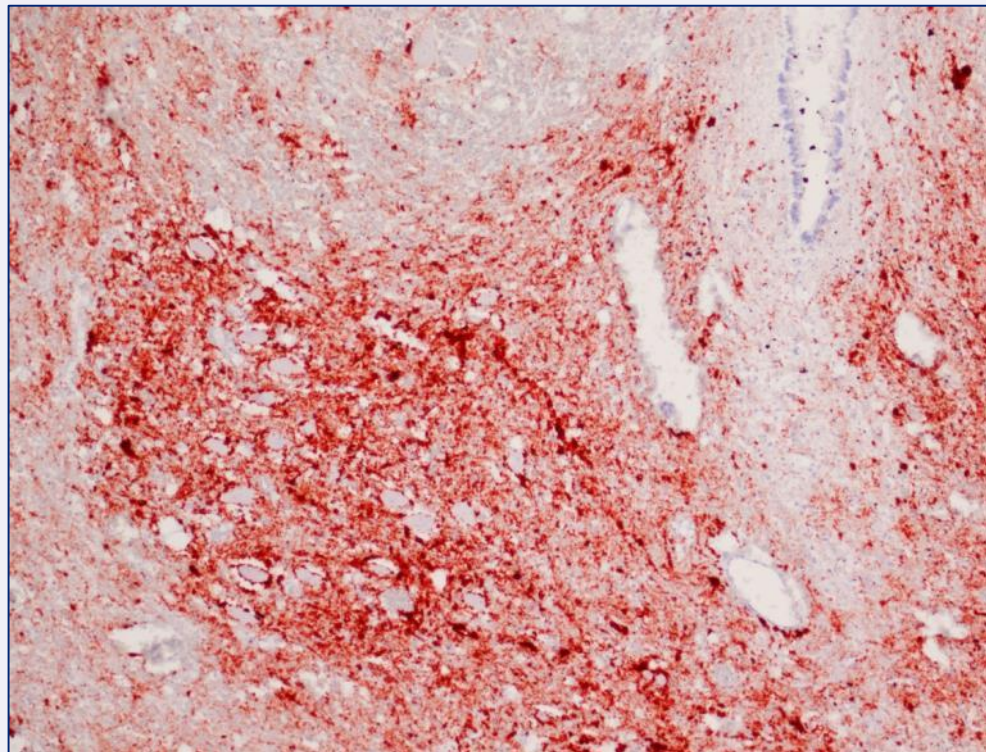
1-Kaleidoscope +MM	
2-16-04-V142 Original	1:20
3-16-04-V142 Medulla	1:20
4-Neg sheep	
5-Nor98 13-50-1873 sheep	1:60
6-Classical scrapie P1213 sheep	1:60
7-16-04-V142 Cerebellum ?	1:20
8-16-04-V142 Midbrain?	1:20
9-16-04-V142 Frontal Cortex	1:40
10-16-04-V142 Frontal Cortex	1:10

WB Linh Tran

Confirmed positive with IHC



IHC Britt Saure



Two new CWD cases, in moose

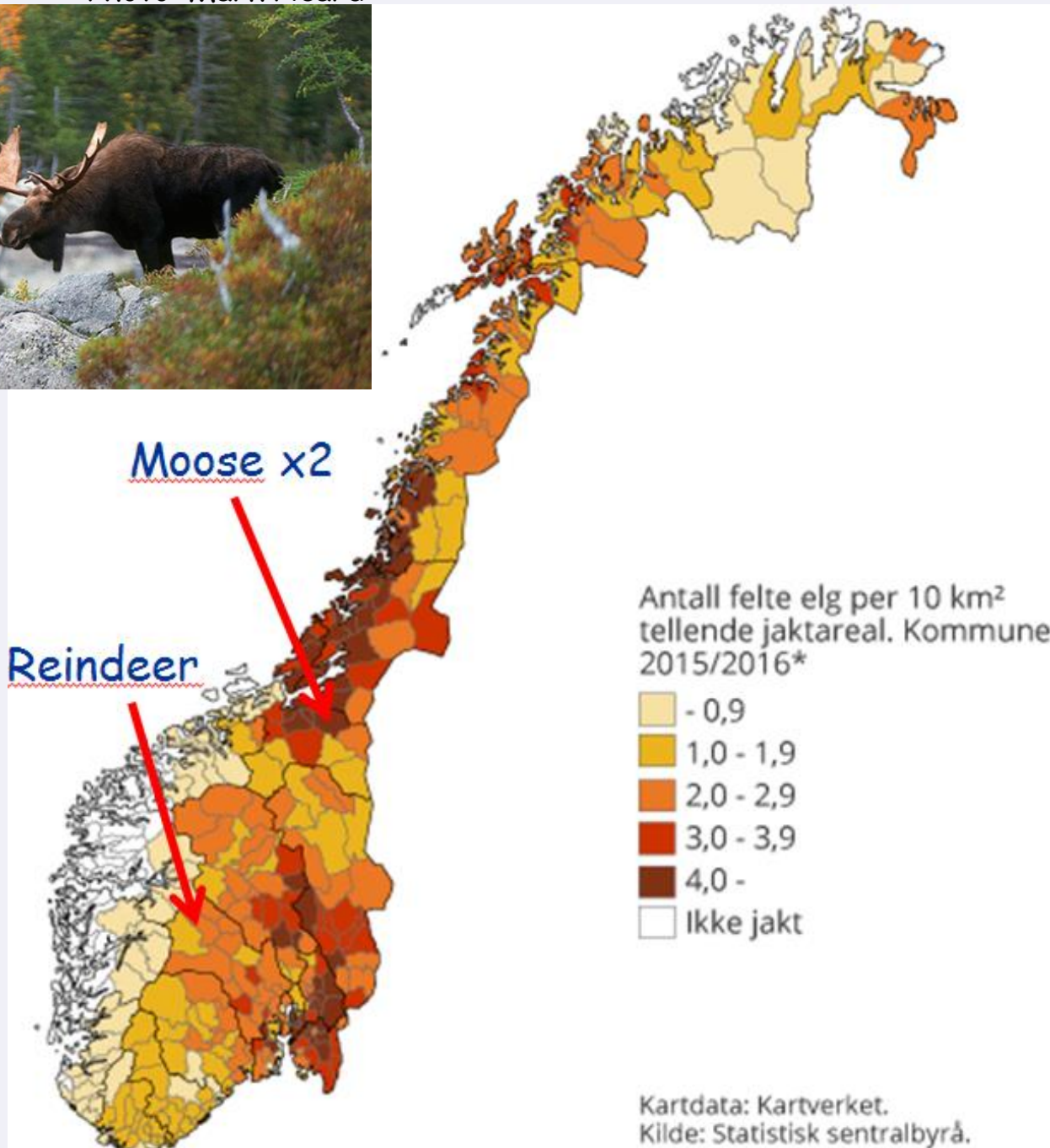


Sent to VI regional laboratory

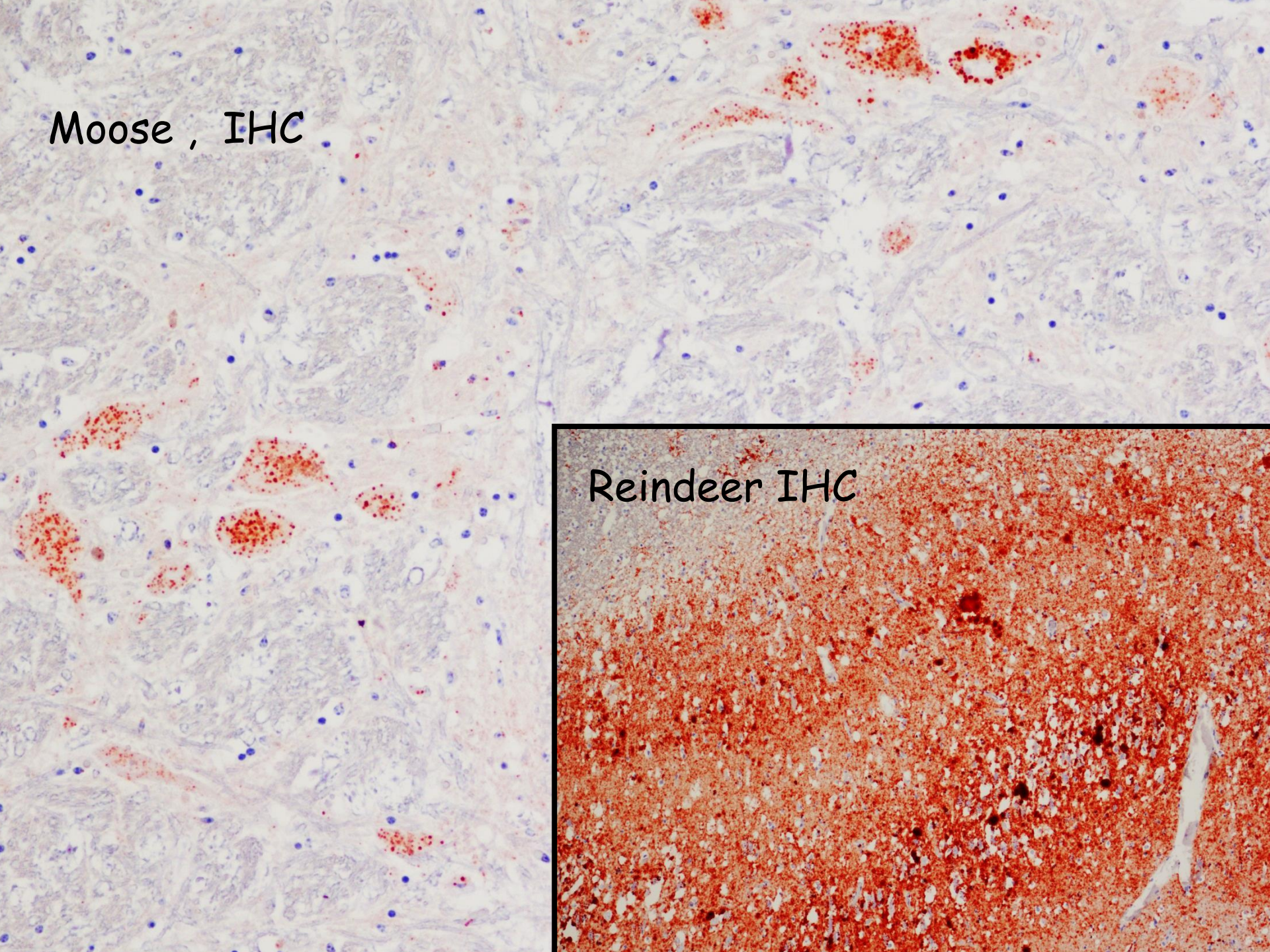
- Moose, pregnant female, emaciated, 13 years old, shot
- Moose, pregnant female, 14 years, found
- dead in a river, trauma, normal condition

Same localisation (Selbu), 320 km from the CWD reindeer's area

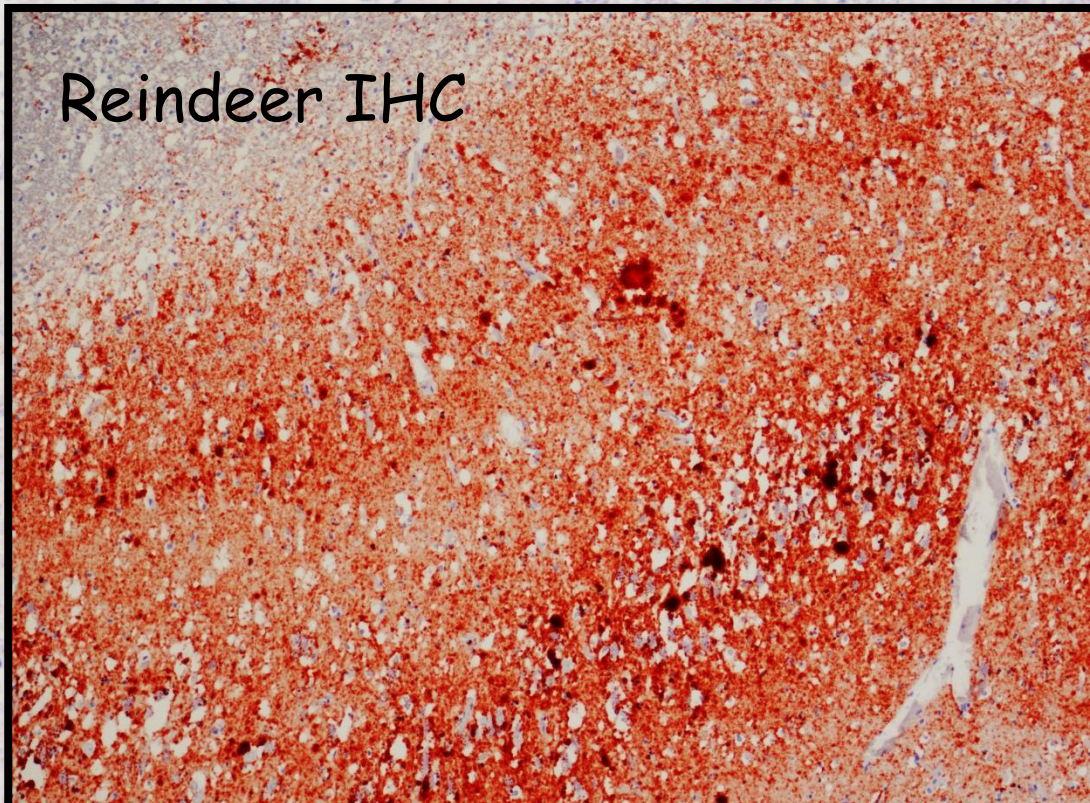
Photo: Mark Picard



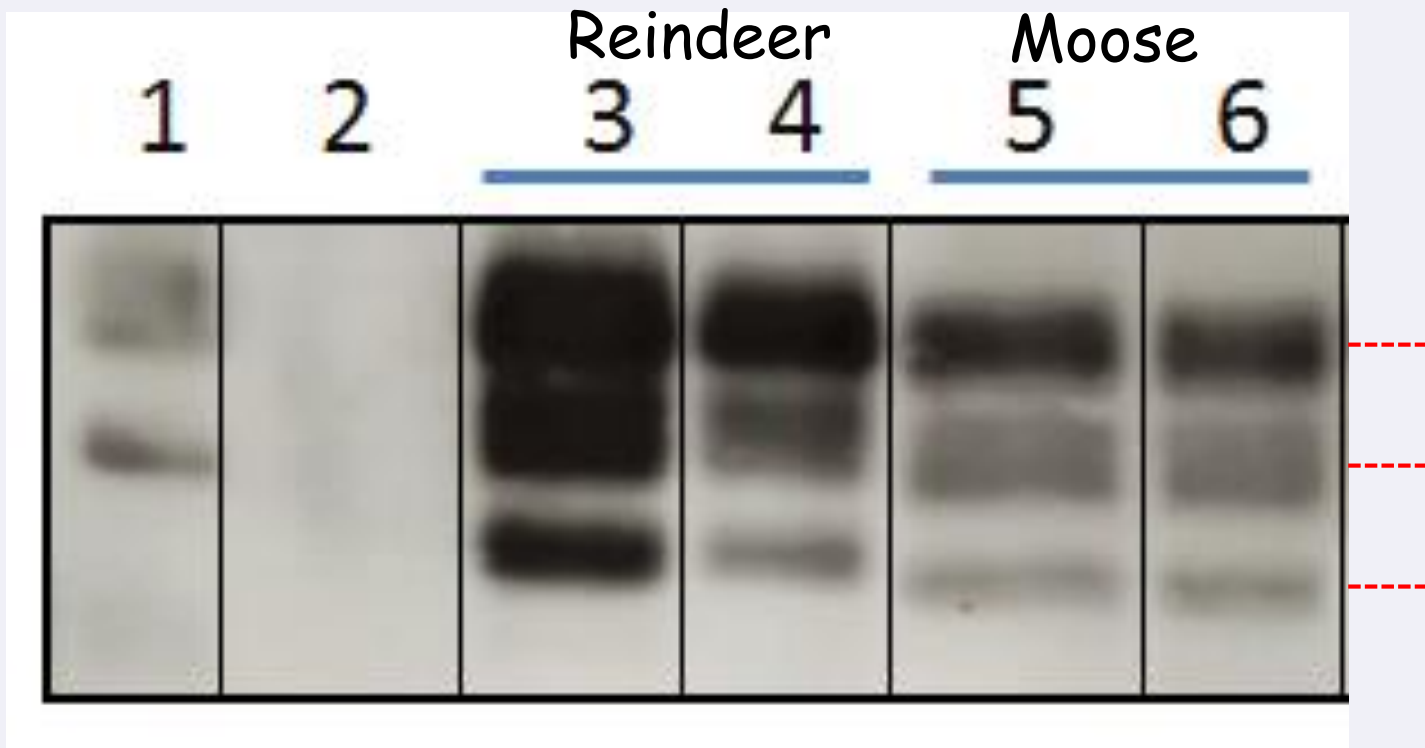
Moose , IHC



Reindeer IHC



Different WB glycosylation pattern






Active surveillance programme in 2016

Category	Analyzed (N)
Moose	4186
Hunted	3777
Unspecified	2
Fallen stock, diseased	407
Deer	2327
Hunted	2068
Farmed slaughtered	83
Unspecified	4
Fallen stock, diseased	172
Reindeer	1635
Hunted	501
Semidomesticated slaughtered	756
Unspecified	11
Fallen stock, diseased	367
Roe deer	307
Hunted	24
Fallen stock, diseased	283
Miscell/Unknown	108
Hunted	90
Fallen stock, diseased	18
Total	8563

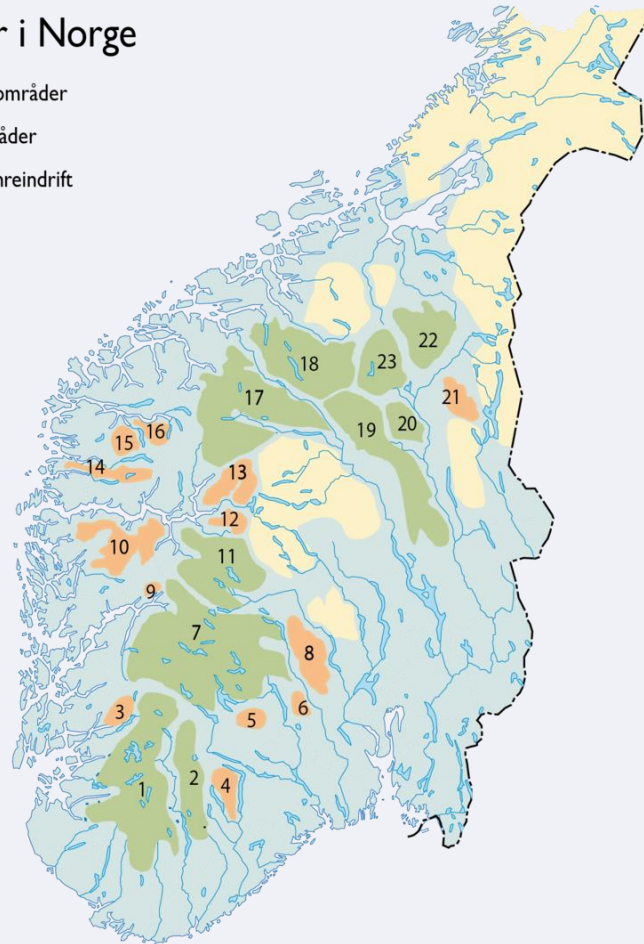


Wild reindeer aeries

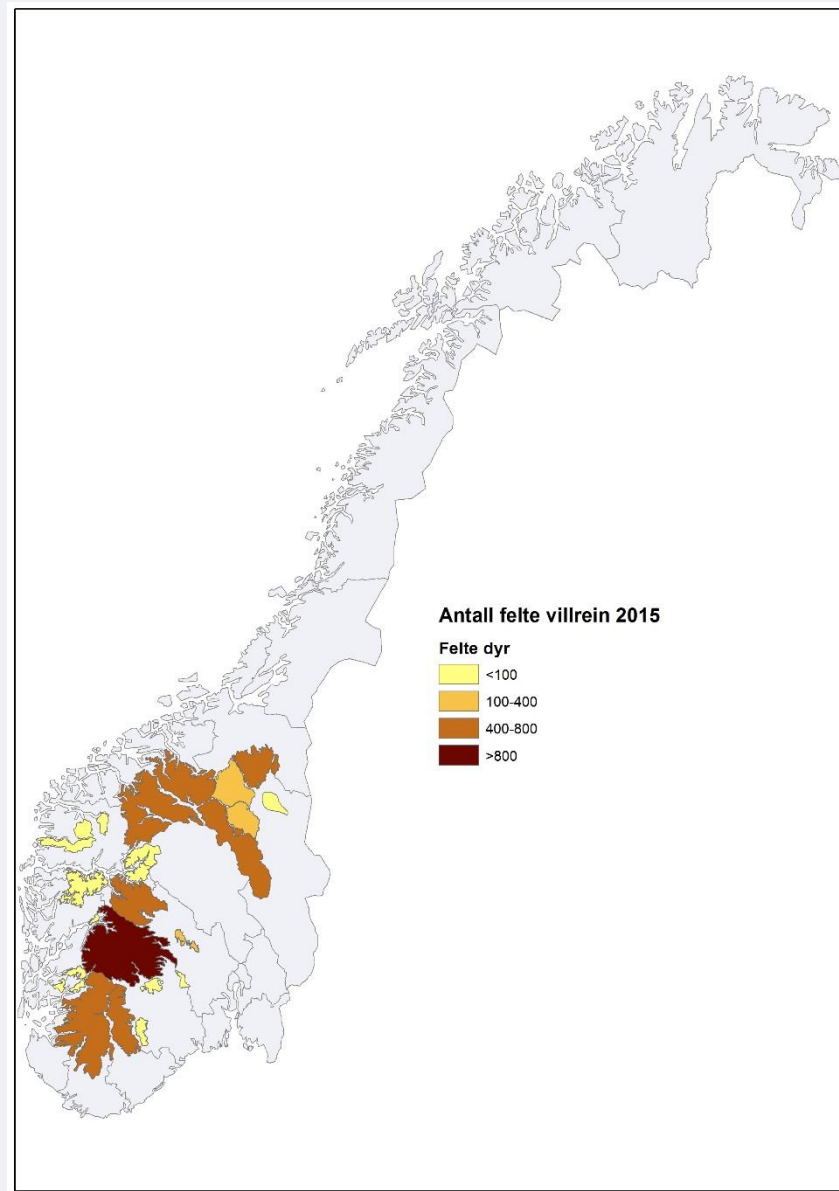
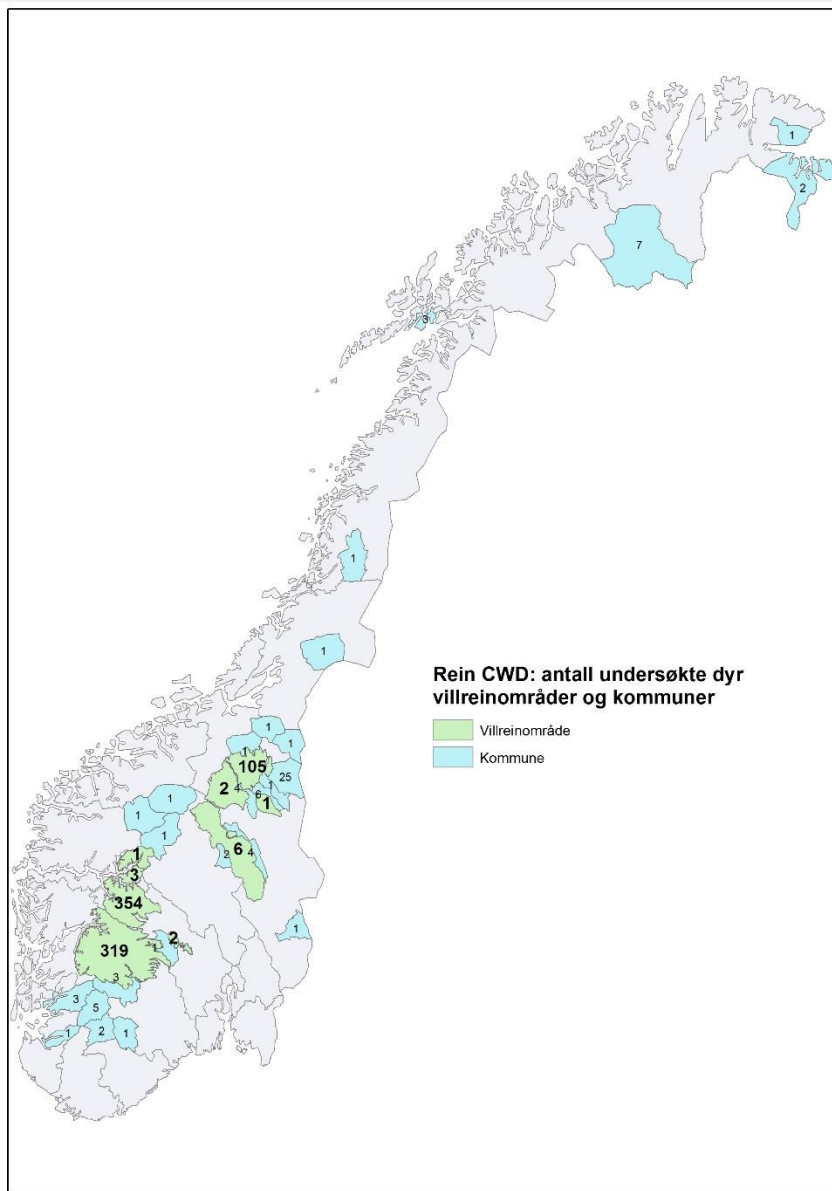
Villreinområder i Norge

-  Nasjonale villreinområder
-  Andre villreinområder
-  Områder med tamreindrift

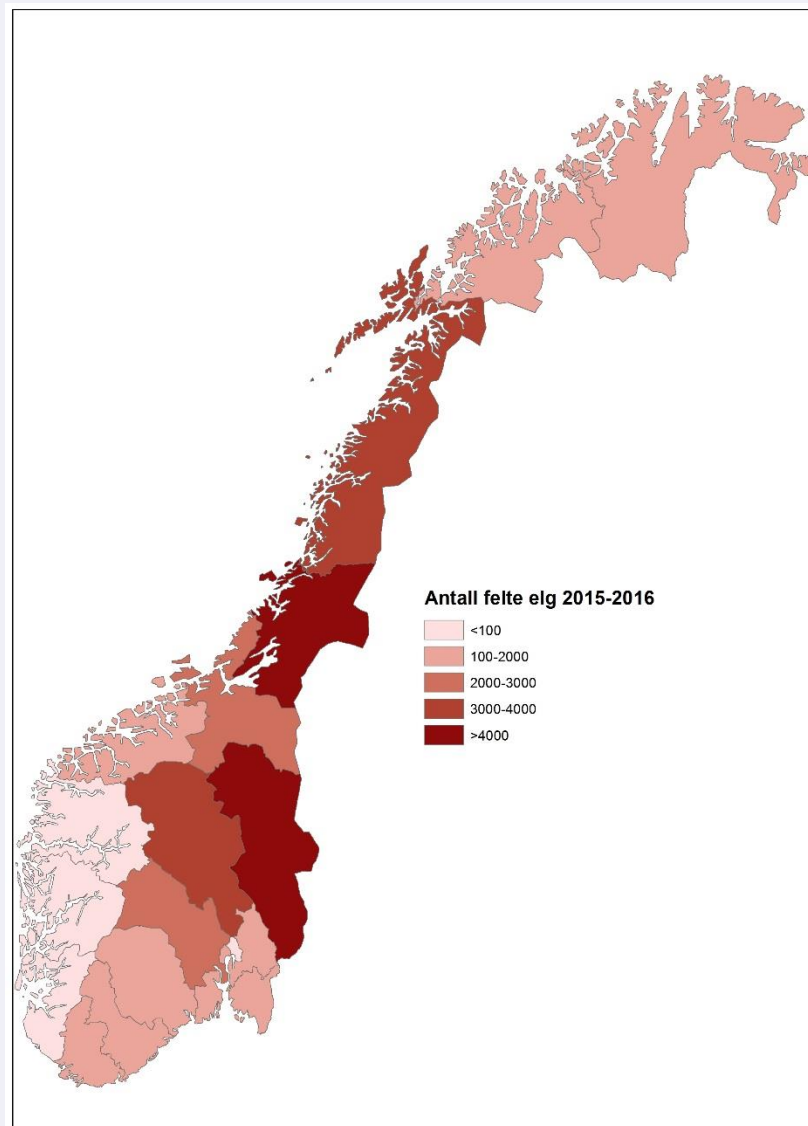
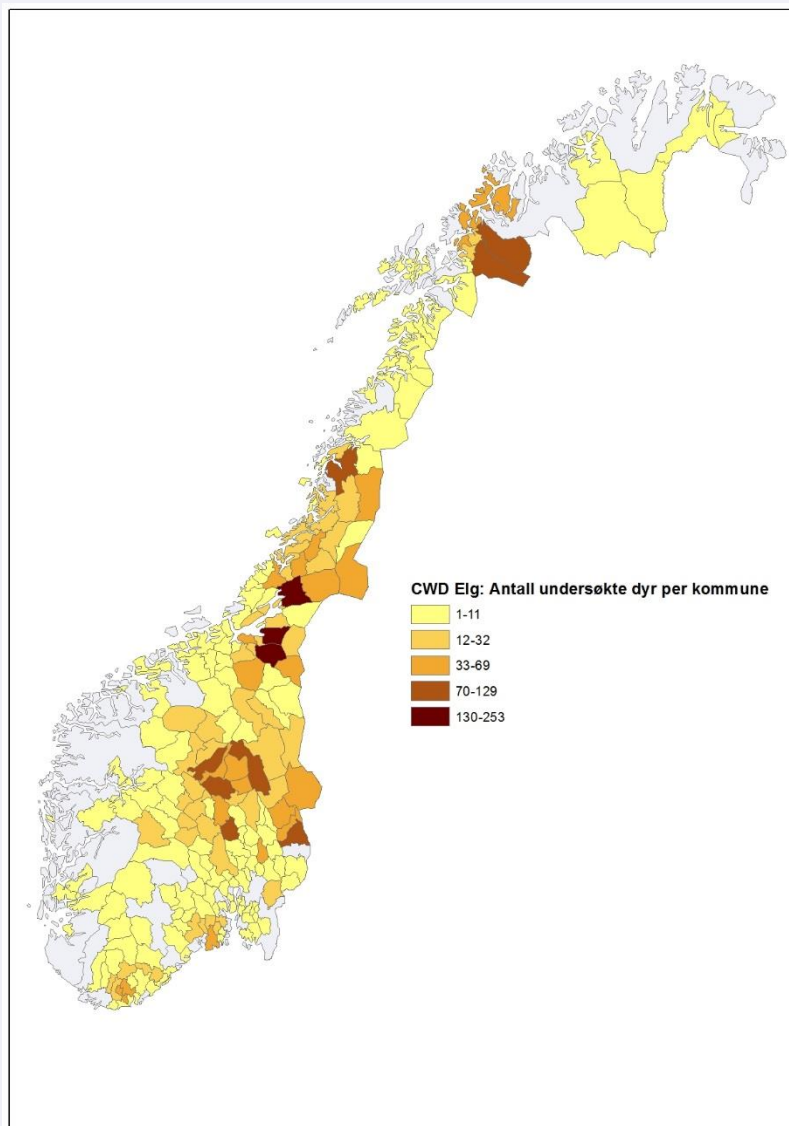
- 1 Setesdal Ryfylke
- 2 Setesdal Austhei
- 3 Skaulen Etnefjell
- 4 Våmur - Roan
- 5 Brattefjell - Vindeggen
- 6 Blefjell
- 7 Hardangervidda
- 8 Norefjell - Reinsjøfjell
- 9 Oksenhalvøya
- 10 Fjellheimen
- 11 Nordfjella
- 12 Lærdal - Årdal
- 13 Vest - Jotunheimen
- 14 Sunnfjord
- 15 Førdefjella
- 16 Svartebotnen
- 17 Reinheimen-Breheimen
- 18 Snøhetta
- 19 Rondane
- 20 Sønkkletten
- 21 Tolga Østfjell
- 22 Forollhogna
- 23 Knutshø



Tested and hunted number of wildliving reindeer

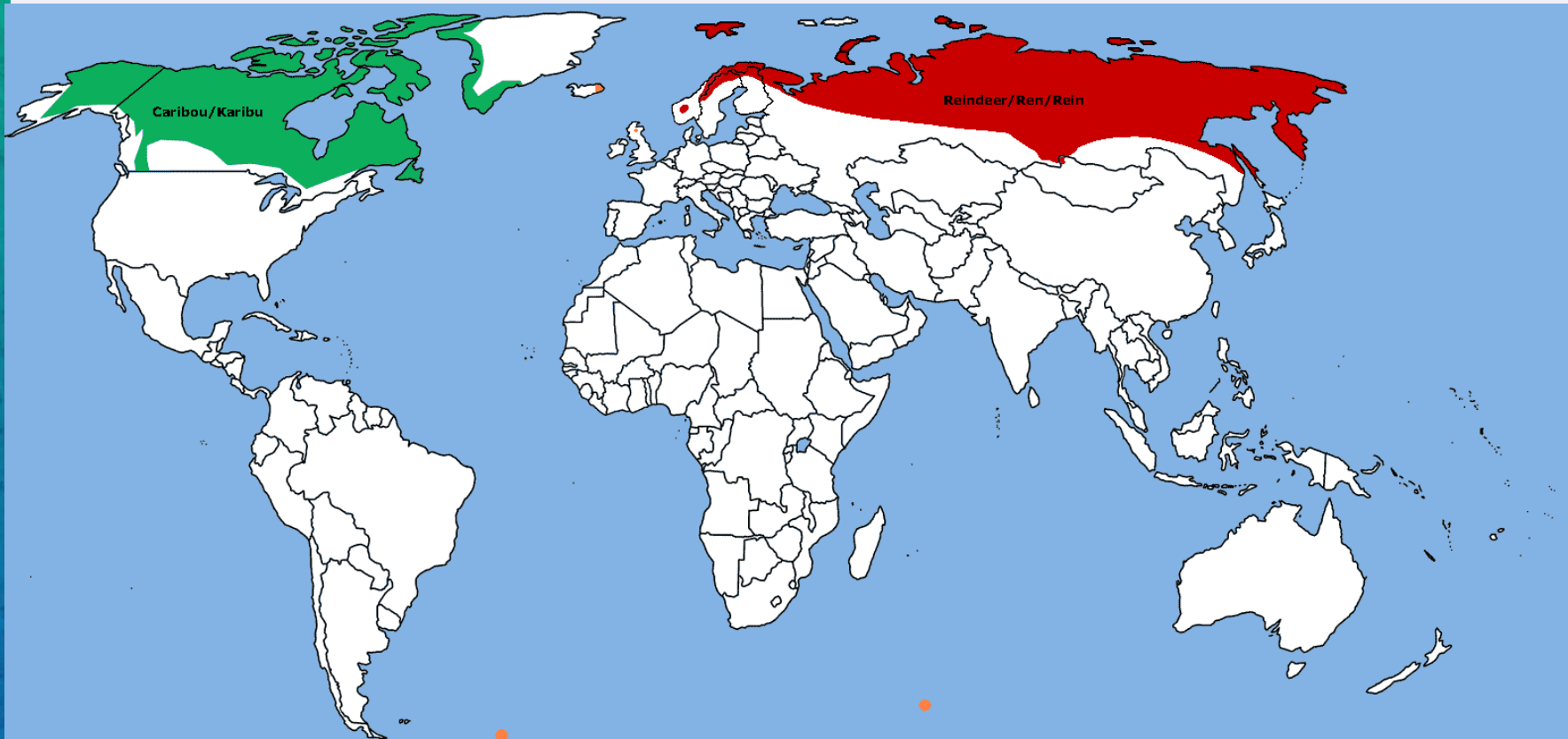


CWD-tested and hunted moose



Questions to be answered:

- Where does it come from
- What is the prevalence and can it be handled?



Where does it come from?

A close-up photograph of a red ink stamp on a piece of light-colored paper. The stamp reads the word "SPECULATION" in a bold, serif, all-caps font. The paper is slightly crumpled and has some circular punch holes visible in the background.

Import of animals

Import food products

From sheep scrapie

From imported urine baits/lures for hunters

Contaminated by a spontaneous case in cervids

Other?

Ongoing research or activity -funded by the Norwegian authorities

- Experimental challenge to mice with different prion types
- Genotyping
- Descriptive pathology
- Diagnostics on fecal materials
- In-vivo diagnostics - gut lymphoids
- Simulation model
- Planning of next year's surveillance

Ongoing experimental challenge trials on transgene mice

Institutions	Country	Recipient	With prion from
INRA Toulouse	France	Transgenic mice	Sheep (3 lines), cattle and human
CEA Jouy-en-Josas	France	Transgenic mice	Macaques
ISS Rome	Italy	Bank voles	
UCL London	UK	Transgenic mice	Human
PRC Fort Collins	USA	Transgenic mice	Cervids and elk
CWRU Cleveland	USA	Transgenic mice	Human
ANSES Lyon	France	Transgenic mice	Sheep
Roslin Edinburgh	UK	Transgenic mice	Human

Thank you for your attention!



A great thank to the involved staff:

Sylvie Benestad, L. Tran, B. Saure, R. Terland, M. Haugum, K. Handeland,
K. Madslien, Ø. Kolbjørnsen, T. Moldal, B. Ytrehus, T. Vikøren



Skrapesjuke

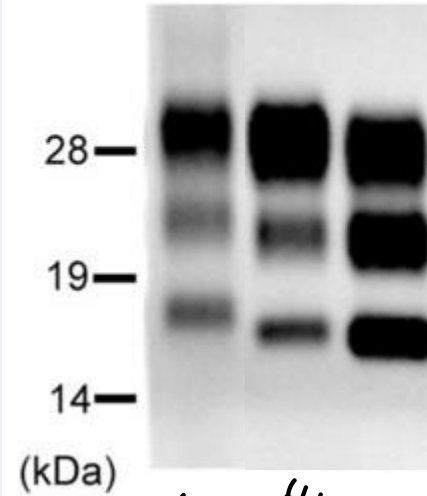


Nor98

Klas Sc

Neg

BSE



28

19

14

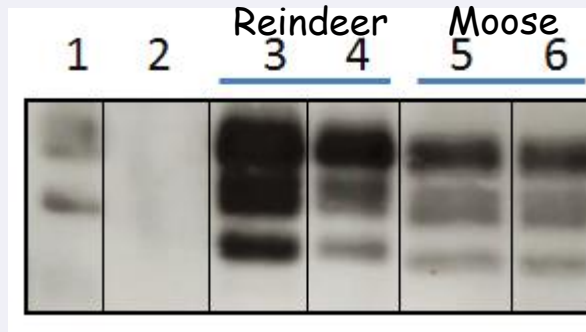
(kDa)

H BSE

Class BSE

L BSE

CWD ???



1

2

Reindeer

Moose

3

4

5

6

mAb: SHa31 and P4



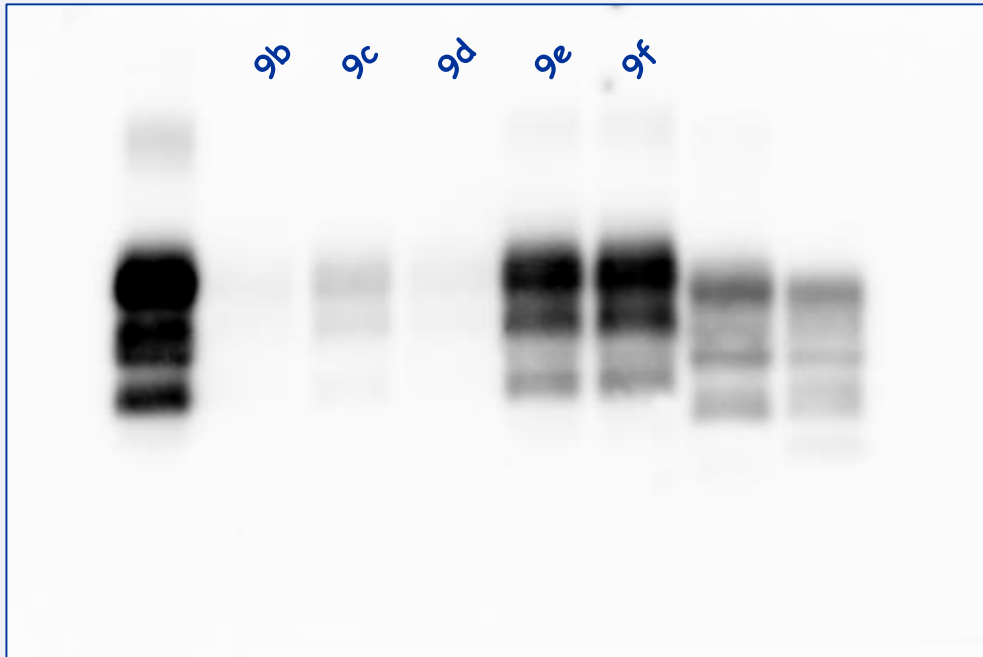
Scrapie

Reindeer-1

Different cortical areas

Moose-1

Moose-2



mAb: L42



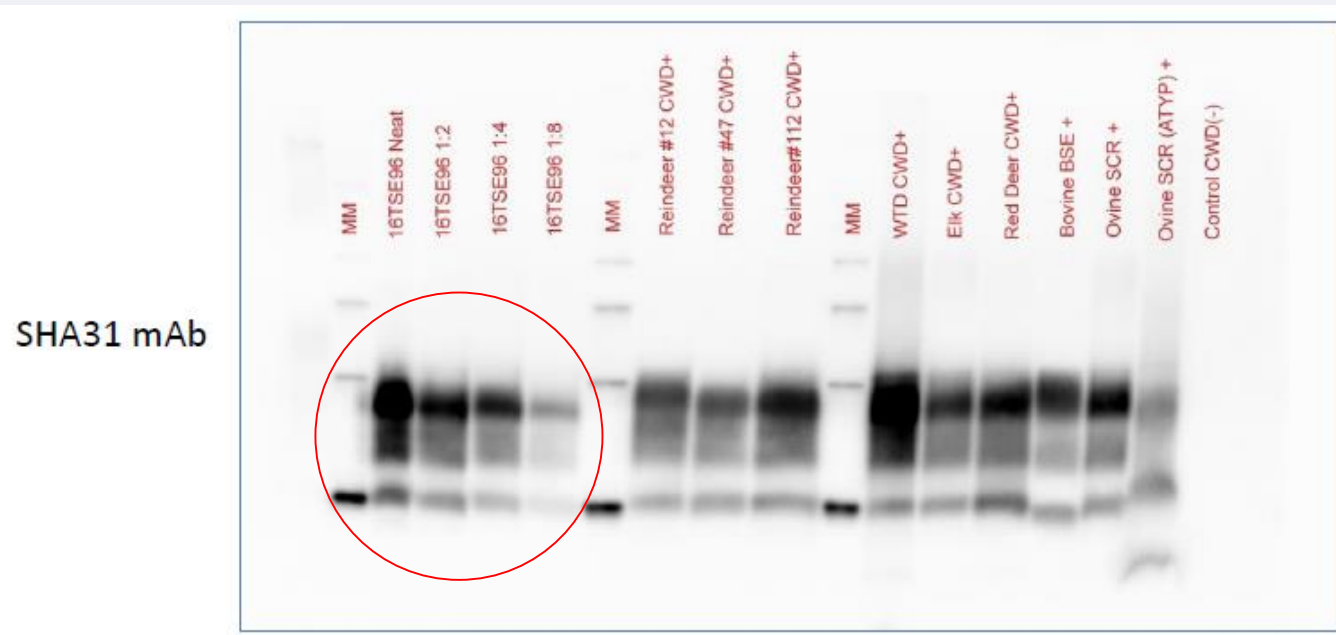
Veterinærinstituttet
National Veterinary Institute

■ Second opinion/ confirmation

from Weybridge
EU ref Lab TSE



and Ottawa
OIE ref CWD



Necropsy findings, *Turid Vikøren*

Female reindeer, not pregnant

> 2,5 years old

Body condition just under average, 43 kg

Multiple hemorrhages and ruptures in the skeletal muscles

General congestion and lung edema



Photos: T. Vikøren

SCIENTIFIC OPINION

Scientific Opinion on the results of the EU survey for Chronic Wasting Disease (CWD) in cervids¹

EFSA Panel on Biological Hazards (BIOHAZ)^{2,3}

European Food Safety Authority (EFSA), Parma, Italy

ABSTRACT

The BIOHAZ Panel was asked to provide a scientific opinion drawing conclusions on the occurrence of CWD in the cervid population in the EU, based on the results of a survey set up by the European Commission and aimed at detecting the possible presence of CWD and other TSEs in wild and farmed cervids in the EU during years 2006 to 2010. The survey was designed taking into account recommendations from an earlier 2004 EFSA opinion and established the minimum sample size to be collected from wild and farmed red deer (*Cervus elaphus elaphus*) from a number of Member States and from wild white-tailed deer (*Odocoileus virginianus*) from Finland. It also required all Member States to collect additional samples from all cervid species. Overall, approximately 13,000 brain stem samples were collected from cervids of different species in 21 Member States and Norway. No TSE positive results were found. The opinion presents, analyses and discusses the results of the survey, explains the uncertainties involved and outlines the limitations of the survey and its results. It is concluded that the lack of one positive TSE test in the farmed and wild red deer and wild white-tailed deer which were sampled indicates that there is not a cervid TSE epidemic in the EU. It is also concluded that, considering the spreading of CWD within and from clusters in North America, the limitations of the sampling performed in the EU CWD/TSEs survey and the known susceptibility of certain cervid species to CWD, occurrence of cases of TSEs, especially in remote and presently unsampled geographic areas, may not be excluded in cervids in the EU. A few recommendations for further experimental studies and possible future monitoring of CWD/TSEs in EU cervids are also provided.

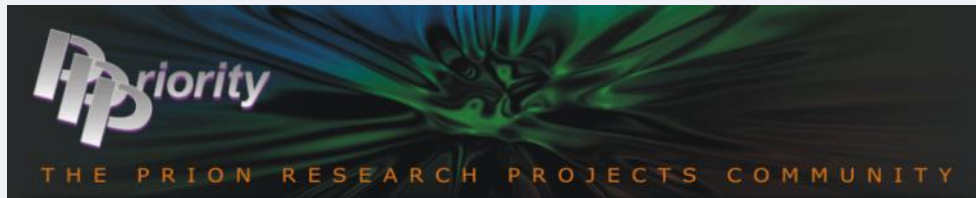
© European Food Safety Authority, 2010

KEY WORDS

Chronic Wasting Disease, Transmissible Spongiform Encephalopathies, monitoring, cervids, red deer, white-tailed deer.



The Priority position paper: protecting Europe's food chain from prions Requena et al., Prion 2016



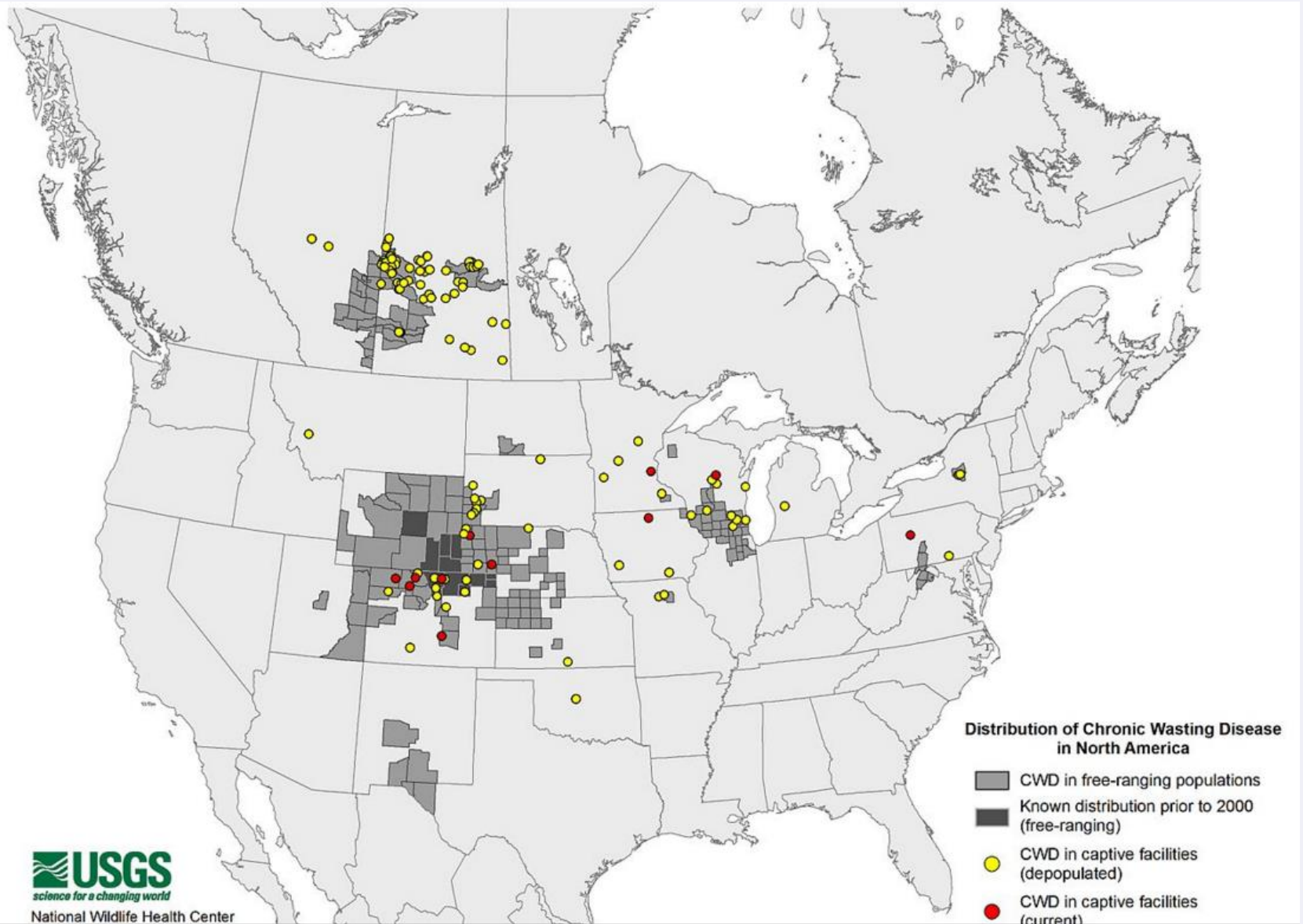
491

492 CWD is a naturally-occurring prion disease in domestic and wild cervids (moose
493 deer and elk), which has reached epidemic proportions in the US and Canada.⁸⁵
494 However, it has not been detected in Europe so far, even though there is significant
495 trafficking of potentially contaminated materials between continents. The potential
496 presence of CWD in Europe is not continuously monitored, although there was a single
497 EU-wide cervid surveillance exercise undertaken some years ago in the context of the
498 EU Regulations for BSE and scrapie, which detected no cases. The deposition of scrapie

499 and CWD prions in the environment occurs through biological fluids and/or faeces.^{39,86}
500 Data depict a scenario where prions may accumulate in the environment due to direct
501 shedding from pre-clinical animals, and remain infectious in soil and water for periods
502 of time long enough to permit transmission to susceptible individuals.^{87,88} Thus CWD,
503 unlike BSE is horizontally and vertically transmitted among captive and wild animals,
504 so it is extremely difficult to eradicate. Given that both red deer and reindeer are native
505 to Europe, there is no reason to assume that CWD could not become a problem in the
506 European continent.



CWD diagnosed in North-America and Korea



Experimental Oral Transmission of Chronic Wasting Disease to Reindeer (*Rangifer tarandus tarandus*)

Gordon B. Mitchell¹, Christina J. Sigurdson^{2,3}, Katherine I. O'Rourke⁴, James Algire¹, Noel P. Harrington¹, Ines Walther¹, Terry R. Spraker⁵, Aru Balachandran^{1*}

1 National and OIE Reference Laboratory for Scrapie and CWD, Canadian Food Inspection Agency, Ottawa Laboratory – Fallowfield, Ottawa, Ontario, Canada, **2** Departments of Pathology and Medicine, University of California, San Diego, La Jolla, California, United States of America, **3** Department of Pathology, Microbiology and Immunology, University of California, Davis, California, United States of America, **4** Animal Disease Research Unit, Agricultural Research Service, United States Department of Agriculture, Pullman, Washington, United States of America, **5** Veterinary Diagnostic Laboratory, Colorado State University, Fort Collins, Colorado, United States of America

Abstract

Chronic wasting disease (CWD), a transmissible spongiform encephalopathy of cervids, remains prevalent in North American elk, white-tailed deer and mule deer. A natural case of CWD in reindeer (*Rangifer tarandus tarandus*) has not been reported despite potential habitat overlap with CWD-infected deer or elk herds. This study investigates the experimental transmission of CWD from elk or white-tailed deer to reindeer by the oral route of inoculation. Ante-mortem testing of the three reindeer exposed to CWD from white-tailed deer identified the accumulation of pathological PrP (PrP^{CWD}) in the recto-anal mucosa associated lymphoid tissue (RAMALT) of two reindeer at 13.4 months post-inoculation. Terminal CWD occurred in the two RAMALT-positive reindeer at 18.5 and 20 months post-inoculation while one other reindeer in the white-tailed deer CWD inoculum group and none of the 3 reindeer exposed to elk CWD developed disease. Tissue distribution analysis of PrP^{CWD} in CWD-affected reindeer revealed widespread deposition in central and peripheral nervous systems, lymphoreticular tissues, the gastrointestinal tract, neuroendocrine tissues and cardiac muscle. Analysis of prion protein gene (*PRNP*) sequences in the 6 reindeer identified polymorphisms at residues 2 (V/M), 129 (G/S), 138 (S/N) and 169 (V/M). These findings demonstrate that (i) a sub-population of reindeer are susceptible to CWD by oral inoculation implicating the potential for transmission to other *Rangifer* species, and (ii) certain reindeer *PRNP* polymorphisms may be protective against CWD infection.

Citation: Mitchell GB, Sigurdson CJ, O'Rourke KI, Algire J, Harrington NP, et al. (2012) Experimental Oral Transmission of Chronic Wasting Disease to Reindeer (*Rangifer tarandus tarandus*). PLoS ONE 7(6): e39055. doi:10.1371/journal.pone.0039055

Editor: Anthony E. Kincaid, Creighton University, United States of America

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Competing Interests: The authors have declared that no competing interests exist.

* E-mail: Aru.Balachandran@inspection.gc.ca





Terry Spraker, CSU, CO

Today,

-52 reindeer (50 free-ranging)

-37 moose

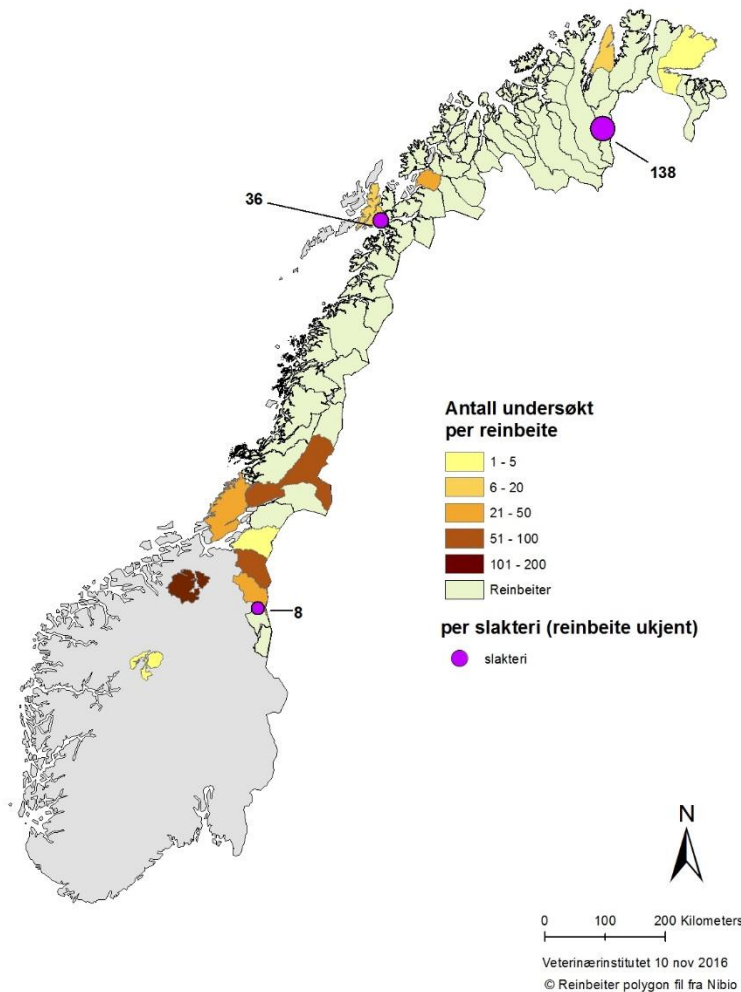
-14 red deer -10 elk

= 113 cervids analysed since the first CWD case

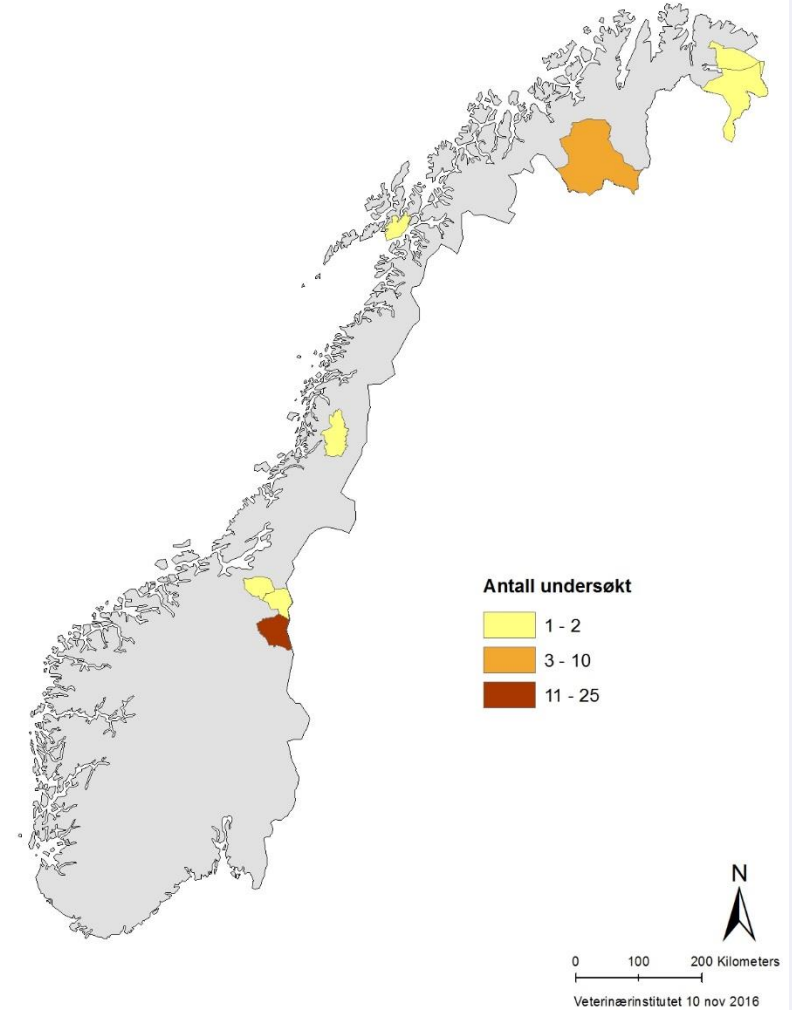
3 positive cases

Tested slaughtered and found dead semidomestic reindeer

Slaktet tamrein undersøkt for CWD



Døde tamrein undersøkt for CWD



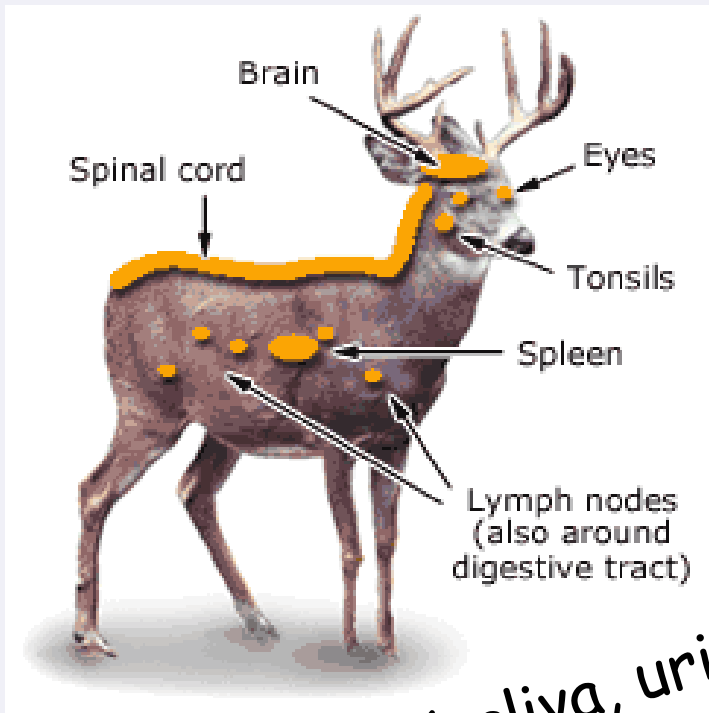
Where does it
come from?

SPECULATION





Nor98 (atypical scrapie) and BSE: prions in the central nervous system



→ CWD = Classical scrapie
. prion distribution in the body
. epidemiology



and excreta (saliva, urine, feces)

The most contagious TSE-disease (direct and via environment)

Measures under consideration / taken



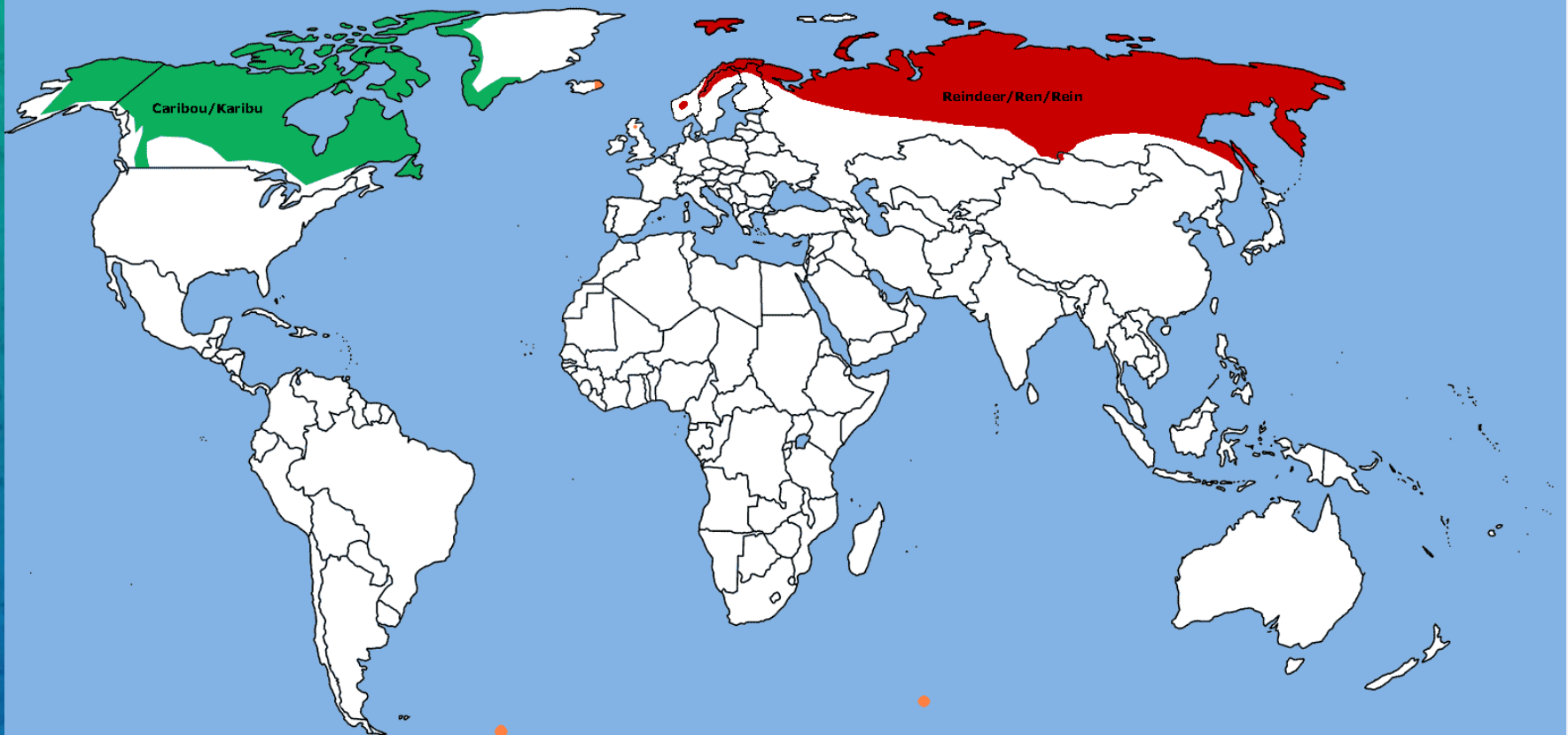
- Norwegian Scientific Committee: risk assessment
- Limited transportation between regions?
- Testing under hunting season
- Testing of fallen stock (whole country, all cervids)
- Testing of slaughtered semi domestic reindeer
- Removal of salt stones?

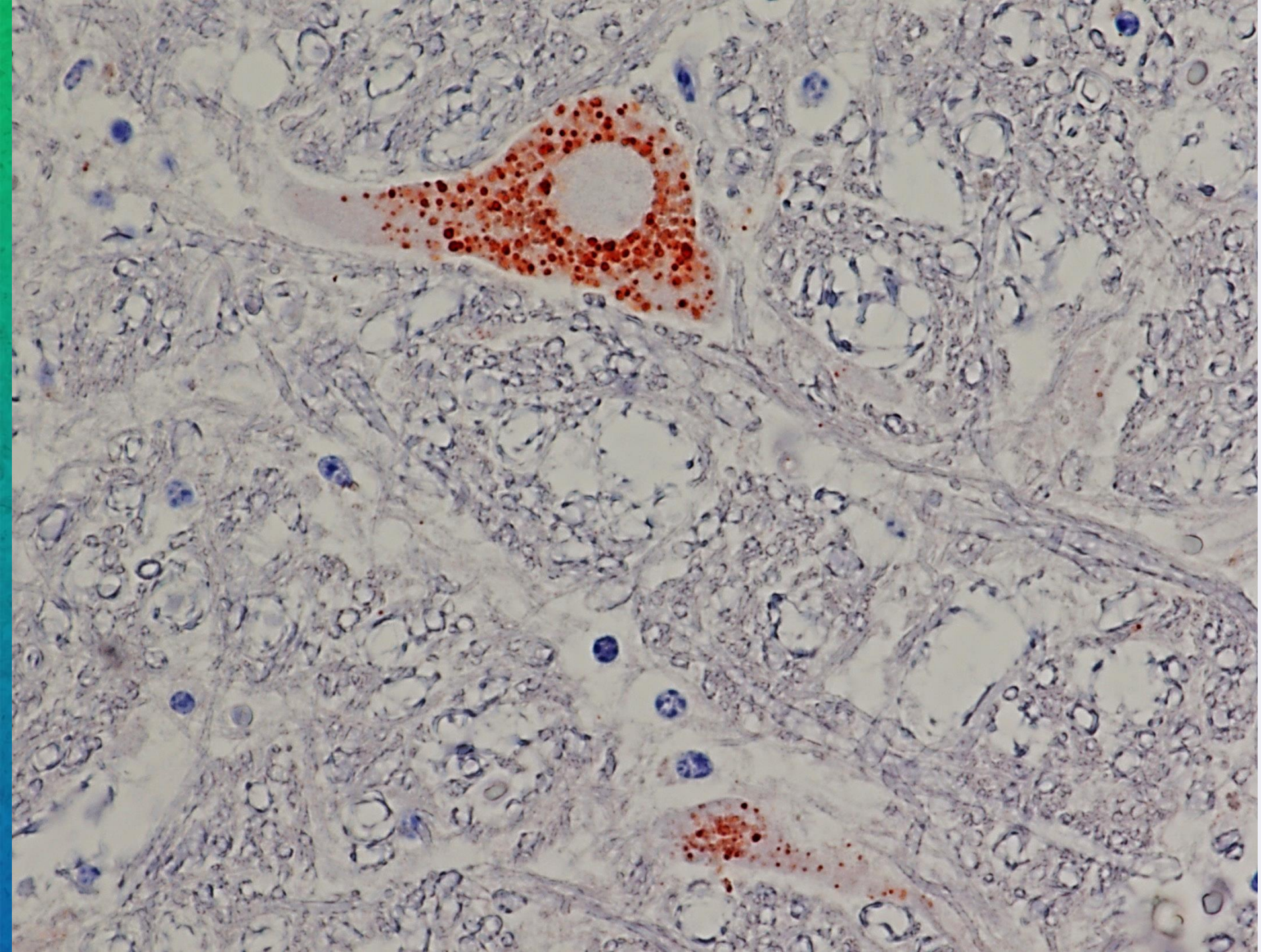
Today, 52 reindeer, 6 moose and 5 red deer (63 cervids) analysed since the first CWD case

3 positive cases

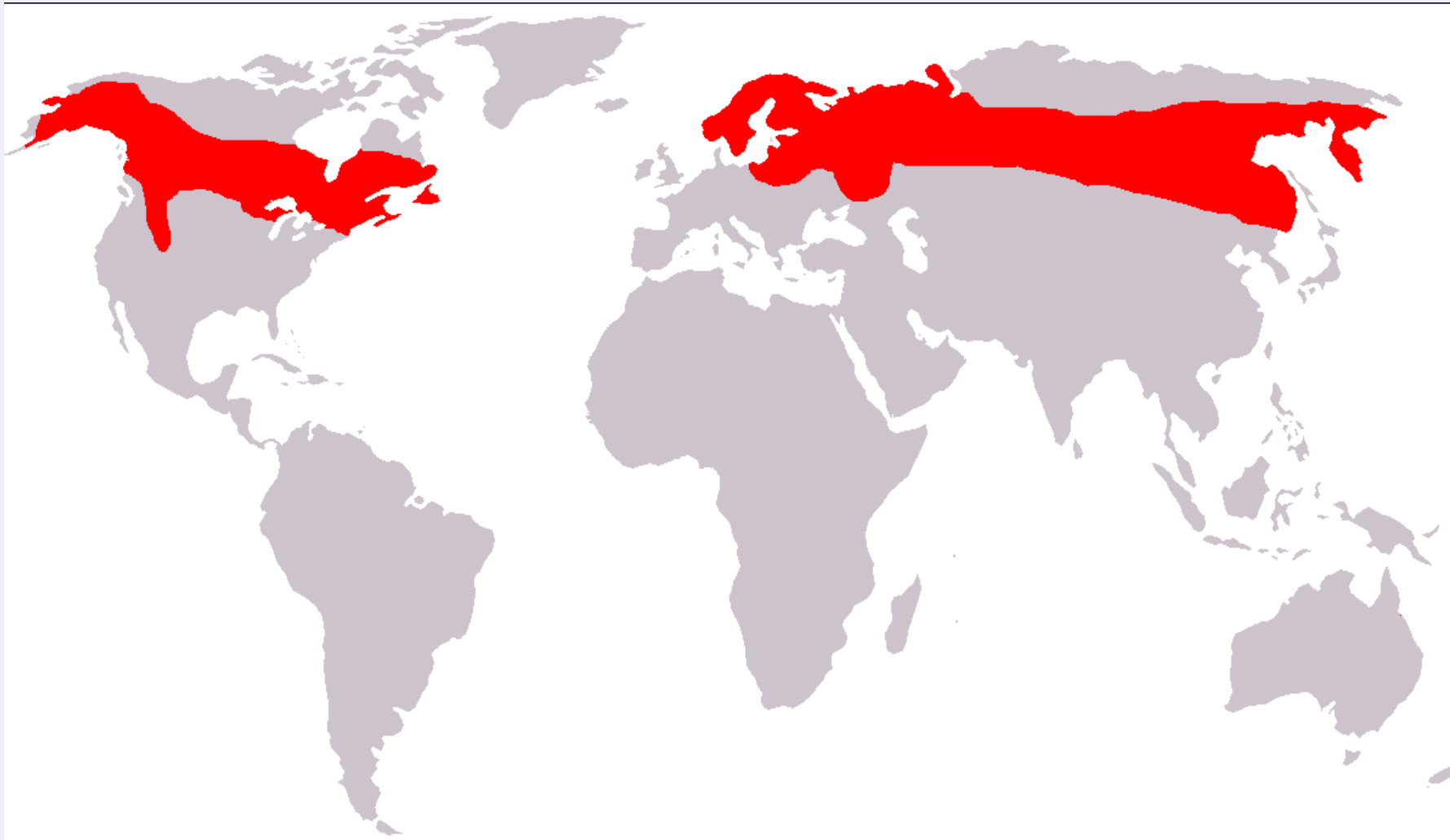
→ Research: characterization of the isolates

Reindeer /Caribou

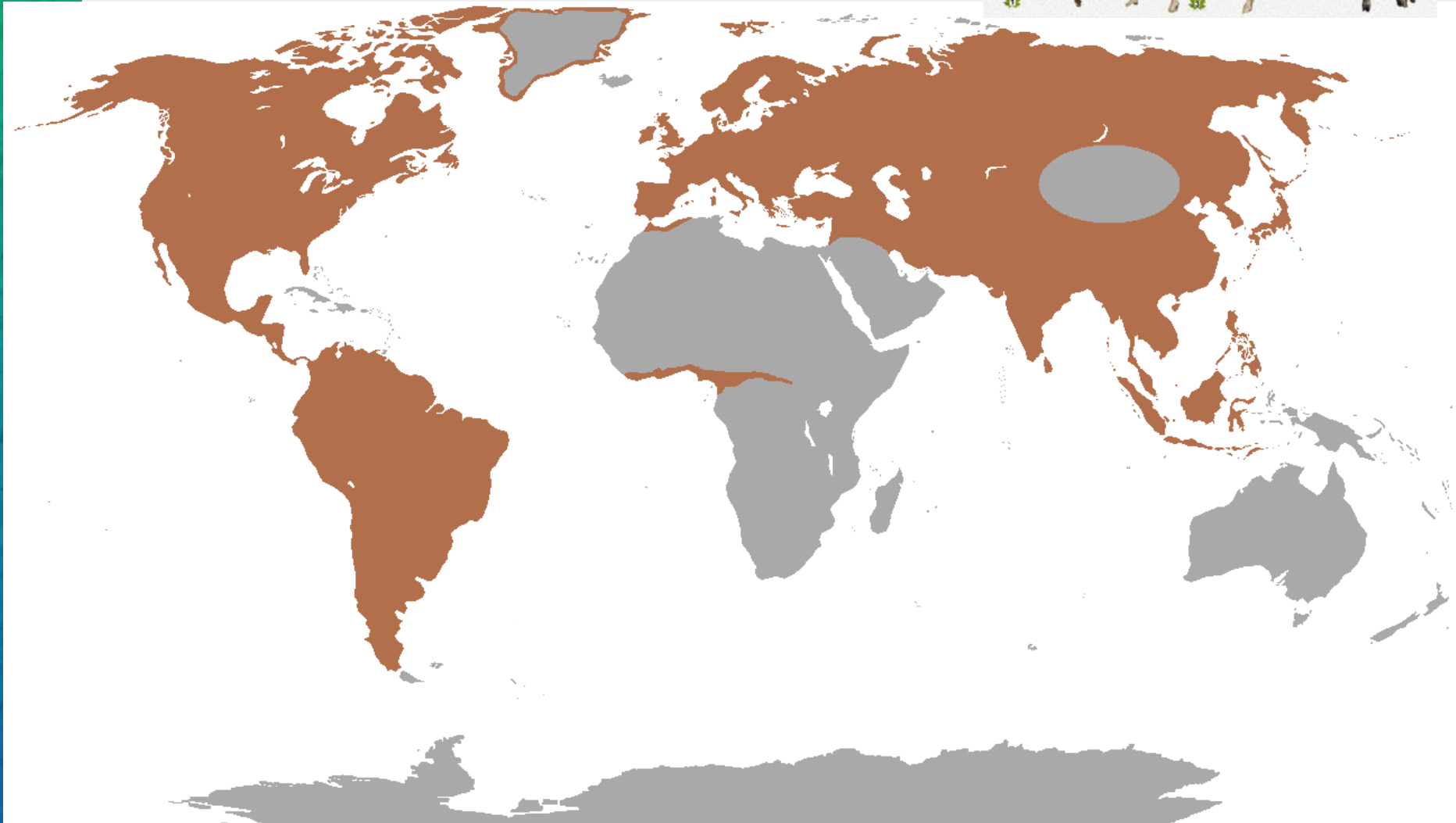




Moose

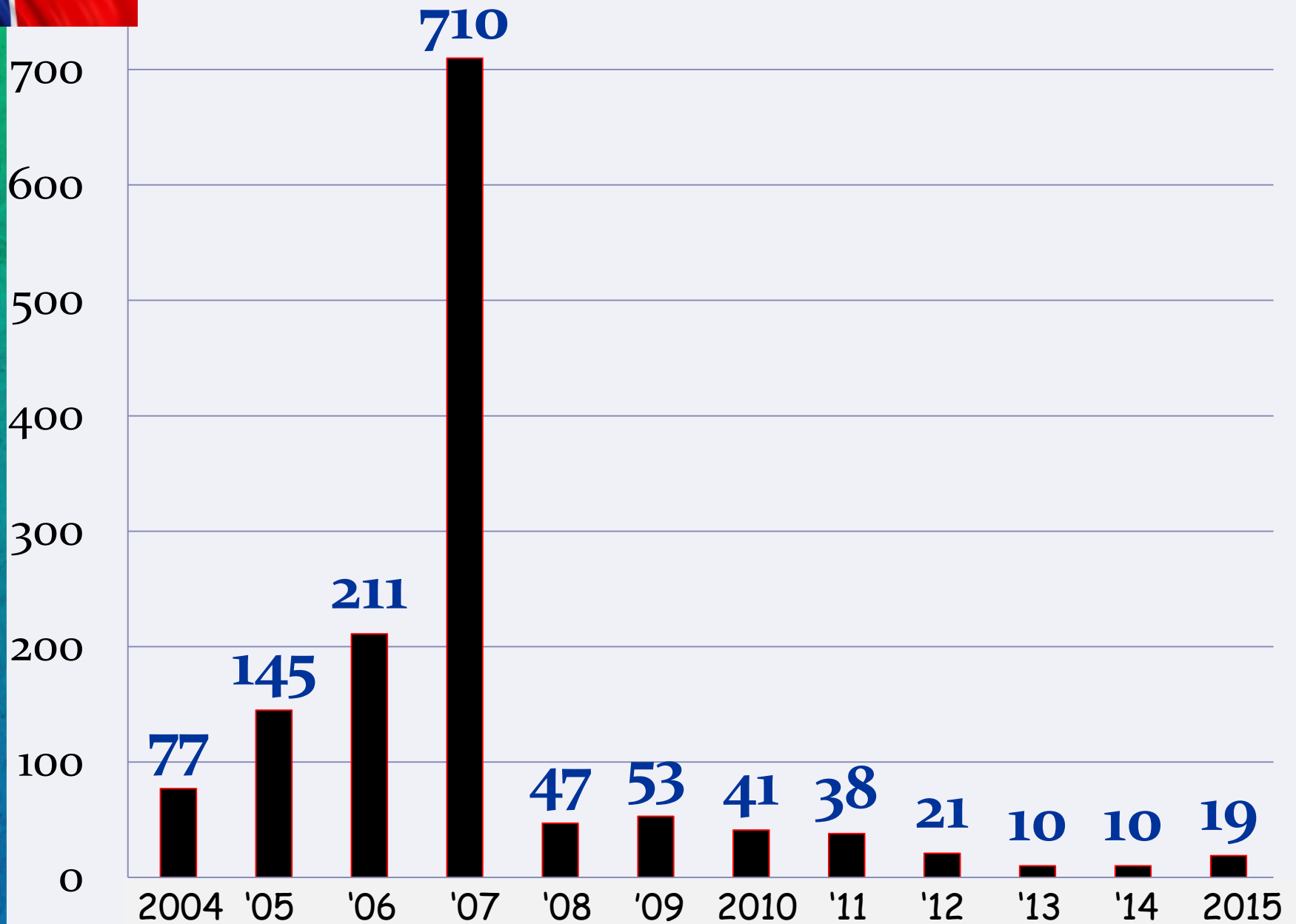


Deer distribution





Analysis of cervids for CWD 2004-2015 *per year*



**CWD
species**

mule deer



white-tailed deer



Moose
(elg)



elk or "wapiti"



CWD clinical signs

Emaciation

Excessive salivation

Behavior changes

Most of the time no signs



Terry Spraker, CSU, CO

Norwegian reindeer (cousin of «caribou»)

