



The surveillance programme for Chronic Wasting Disease (CWD) in free ranging and captive cervids in Norway 2023



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Summary

A total of 14,218 samples of wild, semi-domesticated and captive Norwegian cervids were analysed in 2023. Of these, one moose (*Alces alces*) tested positive for CWD. The case was a 20 years old female killed in Lierne municipality, Trøndelag County.

The case detected had disease characteristics as described for the species in previous reports, i.e. PrP^{Sc} were only detected in brain tissue, and no prions were detected in lymphoid tissues with diagnostic tests. In the Nordic countries CWD have shown sporadic appearance in moose and red deer, whilst in reindeer the disease has been contagious and with lymphoid detections in routine diagnostics.

In addition, 22 wild muskoxen (*Ovibos moschatus*) from the Dovrefjell Mountains were tested and found negative for TSE.

Introduction

Chronic Wasting Disease (CWD) was for the first time detected in Europe in 2016, in Norway [1]. This was also the first detection of a natural CWD infection in reindeer (*Rangifer tarandus*) worldwide. The index case in reindeer was located in Nordfjella management area in Southern Norway, and all subsequent positive reindeer until 2020 were found in this area (Figure 1). The Nordfjella sub-population was eradicated in 2018 [2]. In 2020, one reindeer CWD case was discovered at Hardangervidda, which is the neighbouring area/population to Nordfjella. Another case in reindeer was detected at Hardangervidda in 2022.

CWD is a transmissible spongiform encephalopathy (TSE) or prion disease of cervids [3]. It is an invariably fatal neurodegenerative disease with no known treatment. Well-known in North America, CWD has since the 1960's gradually spread to an increasing number of states and provinces (April 2024: 33 states in USA and five provinces in Canada), both in captive and free-ranging cervids [4]. The republic of Korea has also diagnosed the disease after importation of infected elk (*Cervus elaphus nelsoni*) from Canada [5]. With the disease emergence in Norway, naturally susceptible species also include reindeer. Norway has by now three affected species; reindeer, moose (*Alces alces*) and red deer (*Cervus elaphus*), and in the two latter species CWD has been found in old individuals only. From 2018, CWD has also been diagnosed in Finland and Sweden, in old moose [6].

CWD in the Nordic cervids has shown different epidemiology according to species. Reindeer cases have diagnostic detectable prions both in brain and lymphoid tissues, with moose and red deer cases only in brain, and the geographic distribution suggests a contagious character

of the disease in reindeer. The disease in moose and red deer occurs sporadically and there is little support for contagiousness and horizontal spread between live animals [7].

Four cervid species are prevalent in natural free-ranging populations in Norway: moose, red deer, roe deer (*Capreolus capreolus*) and reindeer. Red deer predominate along the west coast, whereas moose and roe deer mainly inhabit other areas of the country. The wild reindeer is found in fragmented sub-populations in the remote alpine regions of Southern Norway [8]. In addition, Norway has populations of semi-domesticated reindeer that live in a herded condition, though free-ranging. Most semi-domesticated reindeer are found in the northern part of Norway as part of the Sami culture, particularly in the county Finnmark.

The official numbers (March 22, 2024) of hunted cervids were in the 2023-2024 season: 26,007 moose, 52,490 red deer, 32,900 roe deer (numbers per September 1, 2023) and 3,493 free-ranging wild reindeer (numbers per December 15, 2023) [9]. Additionally, the semi-domesticated reindeer population counts about 250,000 animals [10]. There are approximately 90 deer farms in Norway; most of them keep red deer, but some farms have fallow deer (*Dama dama*) and occasionally both species.

Testing wild cervids for CWD was initiated in 2002 through the National Health Surveillance Program for Cervids and muskox (*Ovibos moschatus*), operated by the Norwegian Veterinary Institute (NVI) on behalf of the Norwegian Environmental Agency. From 2003, there has been running a passive surveillance programme for CWD in Norwegian wild and captive cervids, coordinated by NVI and financed by the Norwegian Food Safety Authority. In addition, samples from slaughtered semi-domesticated reindeer from several regions in the country have been tested in the program. In 2006 and 2007 the European Commission (decision 2007/182/EC) initiated a survey for CWD where Norway took part, like the EU member-states, testing 700 cervids. A similar testing was performed in EU in the years 2018-2020 (reg. 2017/1972), covering six member states in addition to Norway. Shown in Table 1 is the total number of cervids tested for CWD in Norway from 2002-2023.

Since there is no separate program to report surveillance of TSE in muskox, such testing is included in this publication. TSE has never been reported in that species.

Table 1: Number and species of cervids tested for CWD in Norway 2002-2023. Additional 41 wild reindeer from Svalbard are not included in the table, all of these from 2018 - 2022 (0 in 2023).

Year	Moose (<i>Alces alces</i>)	Red deer (<i>Cervus elaphus</i>)	Reindeer (<i>Rangifer tarandus</i>)		Roe deer (<i>Capreolus capreolus</i>)	Fallow deer (<i>Dama dama</i>)	Unknown Species	Total
			Semi-domesticated	Wild				
2002-15	142	825	966	10	203	13	0	2 159
2016	4 403	2 582	1 739	842	484	15	87	10 152
2017	5 468	4 083	10 940	2 922	1 955	20	271	25 659
2018	6 705	8 428	12 046	3 650	2 124	48	655	33 656
2019	5 935	5 758	12 937*	3 334	1 692	37	454	30 147
2020	6 200	4 272	6 512	3 213	1 832	92	406	22 527
2021	4 525	4 939	6 141	3 520	1 885	28	623	21 661
2022	3 151	2 641	6 658	3 079	1 817	17	221	17 584
2023	2 026	2 221	5 571	2 176	2 124	5	95	14 218
Total	38 555	35 749	63 510	22 746	14 116	275	2 812	177 763

* Includes two reindeer not specified to wild or semi-domesticated.

Aim

The aim of the programme is to document the occurrence of CWD in the Norwegian cervid populations and TSE in muskoxen.

Materials and methods

The CWD surveillance programme includes testing of slaughtered semi-domesticated reindeer (above 12 months in Southern Norway and above 24 months in Northern Norway), slaughtered farmed deer (above 24 months), euthanized animals and fallen stock of captive deer and semi-domesticated reindeer (above 24 months) and wild cervids (above 12 months), or any animal showing clinical signs, as well as wild cervids submitted for necropsy at the NVI. In addition, since the discovery of CWD in 2016, extensive testing of hunted cervids has been implemented. This effort is a cooperation between the Norwegian Food Safety Authority, the Norwegian Environmental Agency, the Norwegian Institute for Nature Research (NINA), and NVI. Apart from cervids, available wild muskoxen found dead/or killed are also enrolled in the Norwegian surveillance of animal TSE. These animals live in the Dovre Mountains, as the only free-ranging muskox population in Norway.

The routine diagnostics of CWD require testing of brain tissue (*Medulla oblongata*). Due to early detection of prions in lymphoid tissue of reindeer in Norway, such tissues

(retropharyngeal lymph nodes) have, since 2016, been included in the testing of all cervid species and muskoxen when available.

A rapid test (IDEXX HerdChek BSE-Scrapie AG Test, IDEXX Laboratories, Westbrook, USA) was used to screen samples from pooled brain and lymph nodes for detection of PrP^{Sc}.

Initially positive ELISA results were retested in brain and lymph node separately, before confirmatory western-blot (TeSeE® WESTERN-BLOT, Bio-Rad, Marnes-la-Coquette, France), according to the manufacturer's instructions. All the samples were analysed at NVI, being the national reference laboratory for animal TSEs and a WAOH (founded OIE) reference laboratory for CWD.

PRNP variation

PRNP is the gene encoding the cellular prion protein (PrP^C). Variation within this gene is associated to CWD susceptibility and development [11,12]. In Norwegian cervids *PRNP* variation differs between species, reindeer being most polymorphic, followed by red deer, moose and roe deer respectively. The latter being monomorphic [13]. All animals detected positive for CWD in the current surveillance programme have been sequenced and *PRNP* variation has been recorded (Table 2). The *PRNP* allele nomenclature/genotype presented in this report follows Güere et al. [12,13]

Table 2: Genotype variation in Norwegian cervids diagnosed with CWD. For each species, the genotype nomenclature is in accordance with Güere et al. [12,13].

Year	Reindeer				Moose			Red deer
	A/A	A/B	A/C	A/D	B/C	109KK	109QQ	226EE
2016	2		2			2		
2017	5	3			1	1		1
2018	3	1	2				1	
2019						1	1	
2020				1		1		
2021						2		1
2022	1					1	1	1
2023						1		
Total	11	4	4	1	1	9	3	3

Results

In total, samples from 14,218 individual cervids were analysed in 2023 (Table 1), of which one wild moose tested positive for PrP^{Sc}. This animal shared diagnostic characteristics previously described for Nordic moose [14,15], with diagnostic detectable prions only in the brain tissue.

Semi-domesticated reindeer (5,571) contributed with about 39% of the total of analysed cervids. Moose samples counted 2,026 and red deer 2,221, being the two larger subgroups beside slaughtered semi-domesticated reindeer. The number of tested roe deer, fallow deer and wild reindeer, was 2,124, 5 and 2,176, respectively. Amongst the total number were 95 animals of unknown species. In addition, 22 muskoxen were tested and found negative.

A total of 73% of the animals were tested by analysing both lymphoid tissue and brain. From the remaining animals only brain or lymphoid tissue had been received and from 26% of the animals only material from brain were tested and from less than 0.5% of the animals only material from lymphoid tissue were tested. Table 3 and Figures 2-9 give the numbers, species and geographical distribution of cervids tested for CWD in 2023.

Table 3: Number of cervids tested in the Norwegian surveillance programme for CWD 2023, distributed on species and reason for submission. Additional 22 muskoxen are not included in the table.

Species	Wild			Captive and semi-domesticated			Unknown	Total
	Hunted	Diseased, injured or traffic killed	Unknown	Slaughtered	Diseased, injured or traffic killed	Unknown		
Moose	983	702	341	0	0	0	0	2 026
Red deer	1 277	324	342	271	6	0	1	2 221
Reindeer	1 780	32	364	5 459	109	3	0	7 747
Roe deer	49	1 831	244	0	0	0	0	2 124
Fallow deer	0	0	1	3	1	0	0	5
Unknown	5	3	67	18	3	0	1	95
Total	4 094	2 892	1 359	5 751	117	3	2	14 218

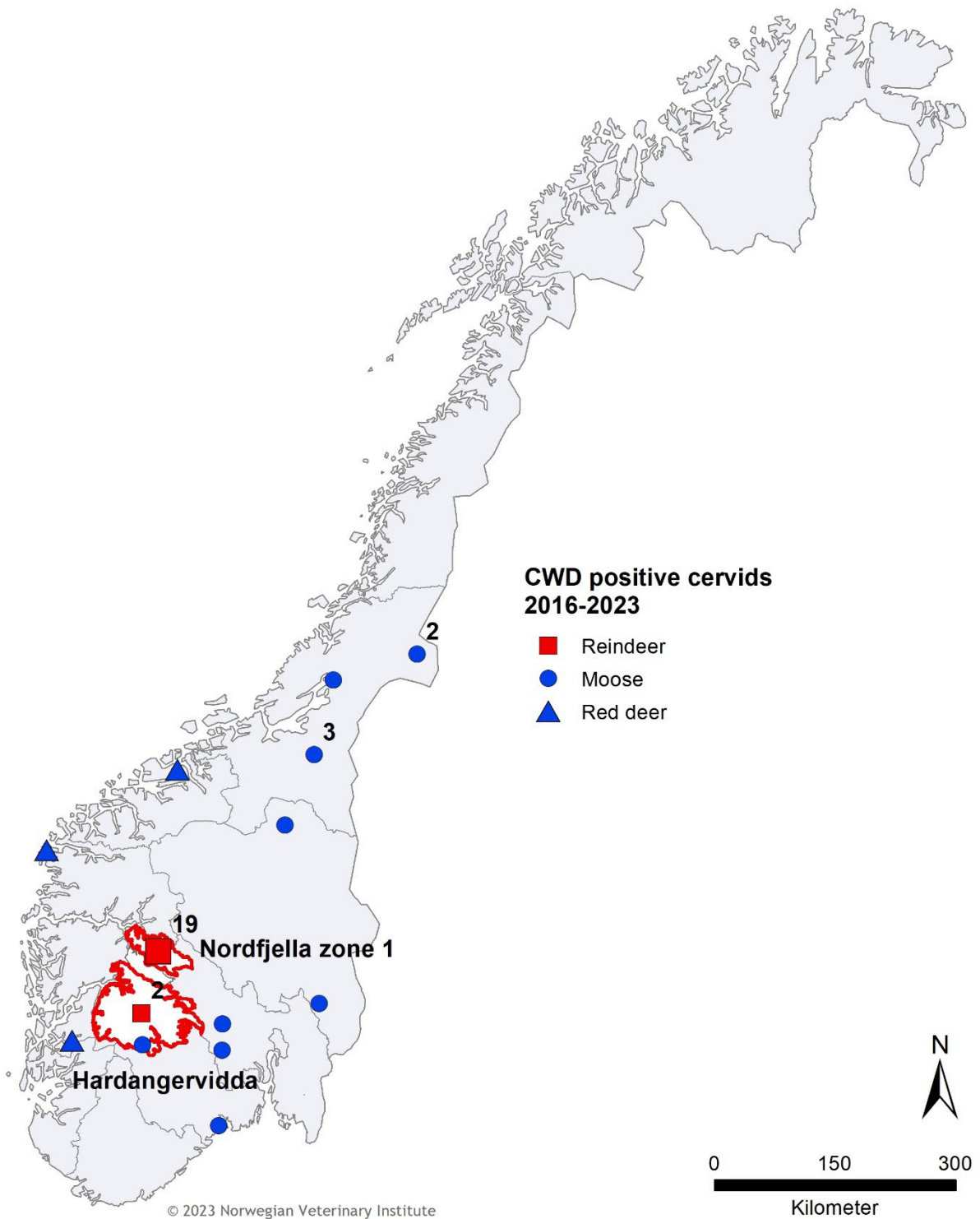


Figure 1: Geographical location of Nordfjella and Hardangervidda (encircled in red), and municipalities in which the total number of CWD positive cervids have been detected through the Norwegian surveillance programme for CWD. Unless a single case, the number of animals at each location is given.

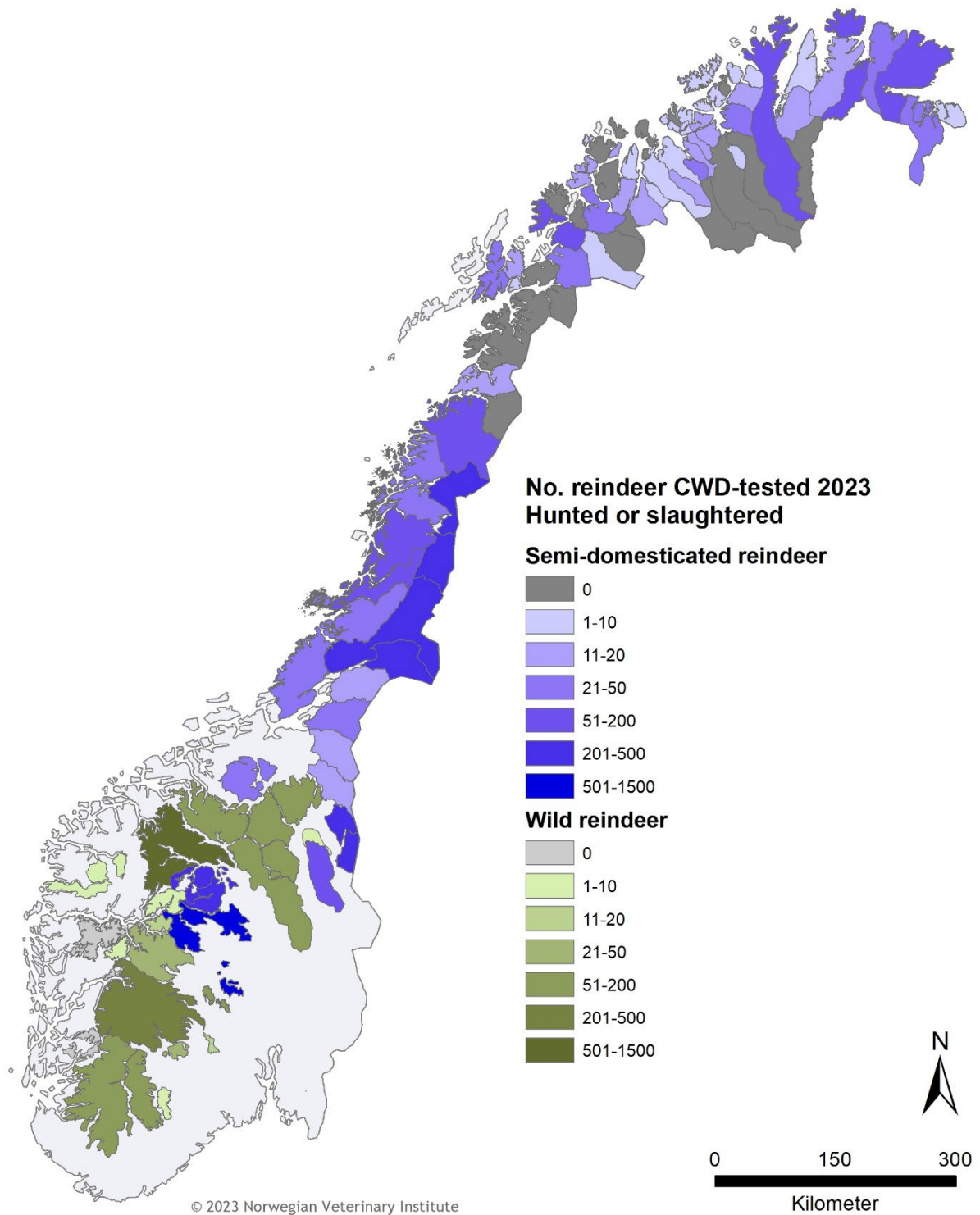


Figure 2: Number and geographical distribution of hunted free-ranging (green) and slaughtered semi-domesticated (blue) reindeer (*Rangifer tarandus*) tested in the Norwegian surveillance programme for CWD in 2023.

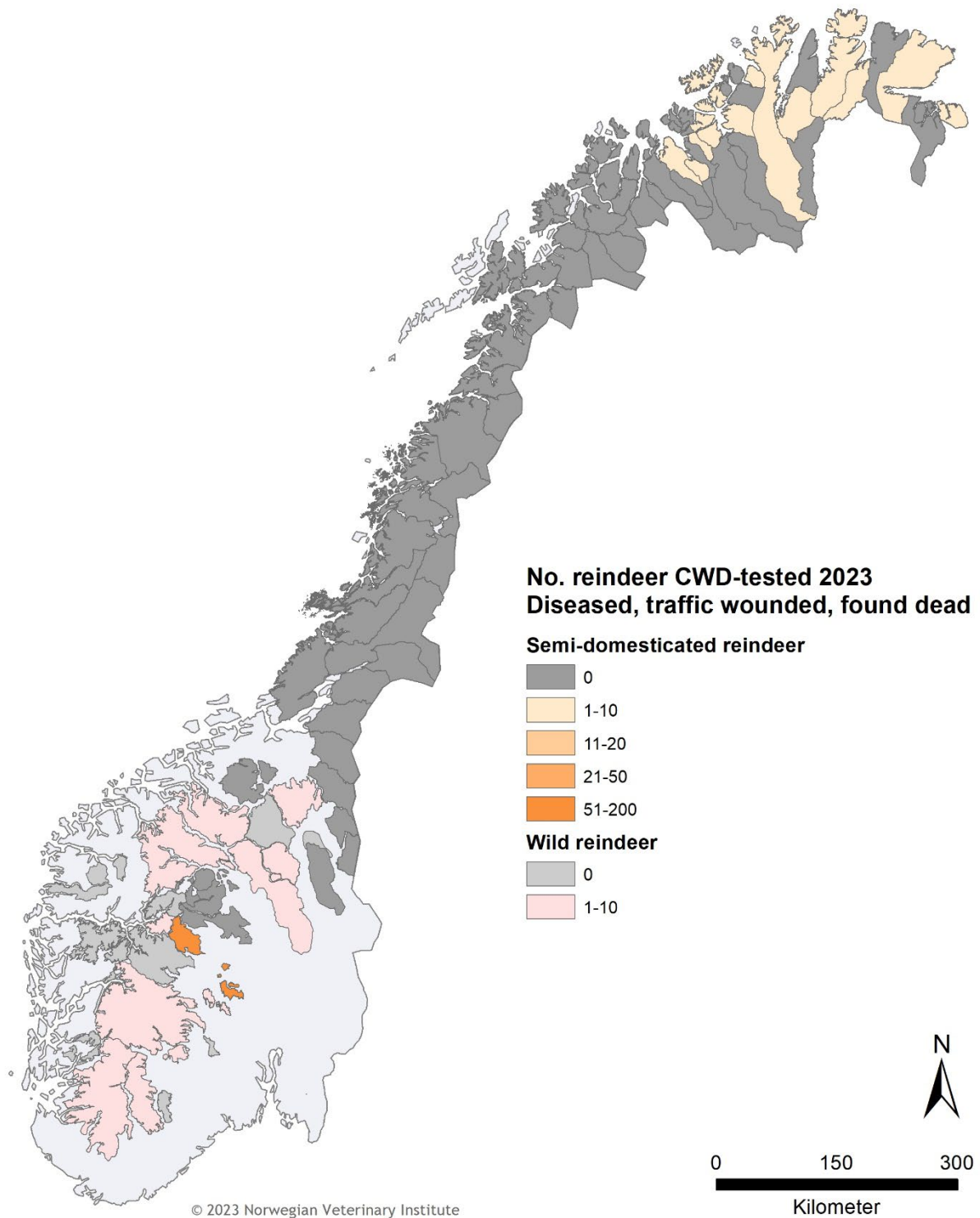


Figure 3. Number and geographical distribution of reindeer (*Rangifer tarandus*), both free-ranging and semi-domesticated, found diseased and euthanised, traffic wounded or dead and tested in the Norwegian surveillance programme for CWD in 2023.

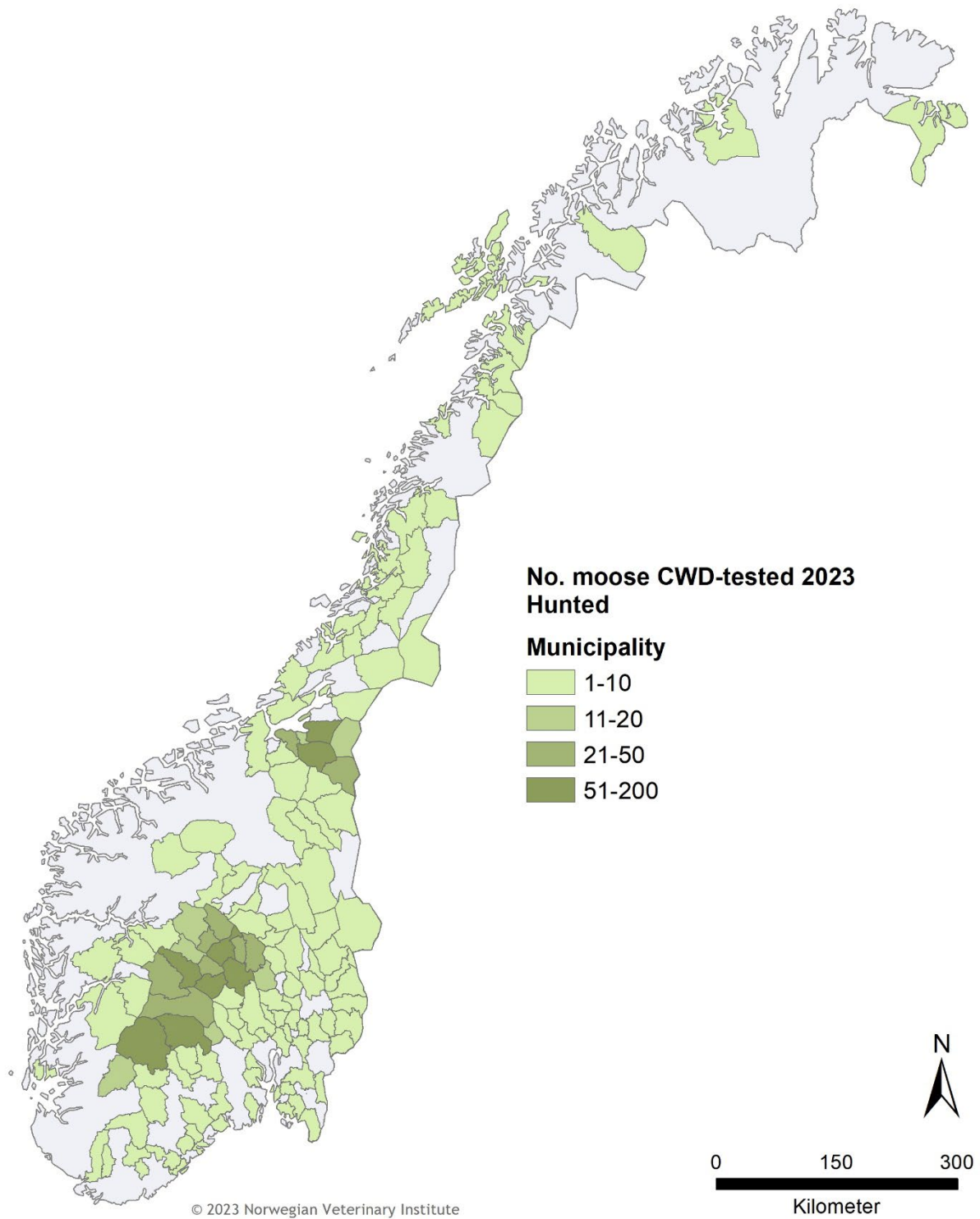


Figure 4: Number and geographical distribution of hunted free-ranging moose (*Alces alces*) tested in the Norwegian surveillance programme for CWD in 2023.

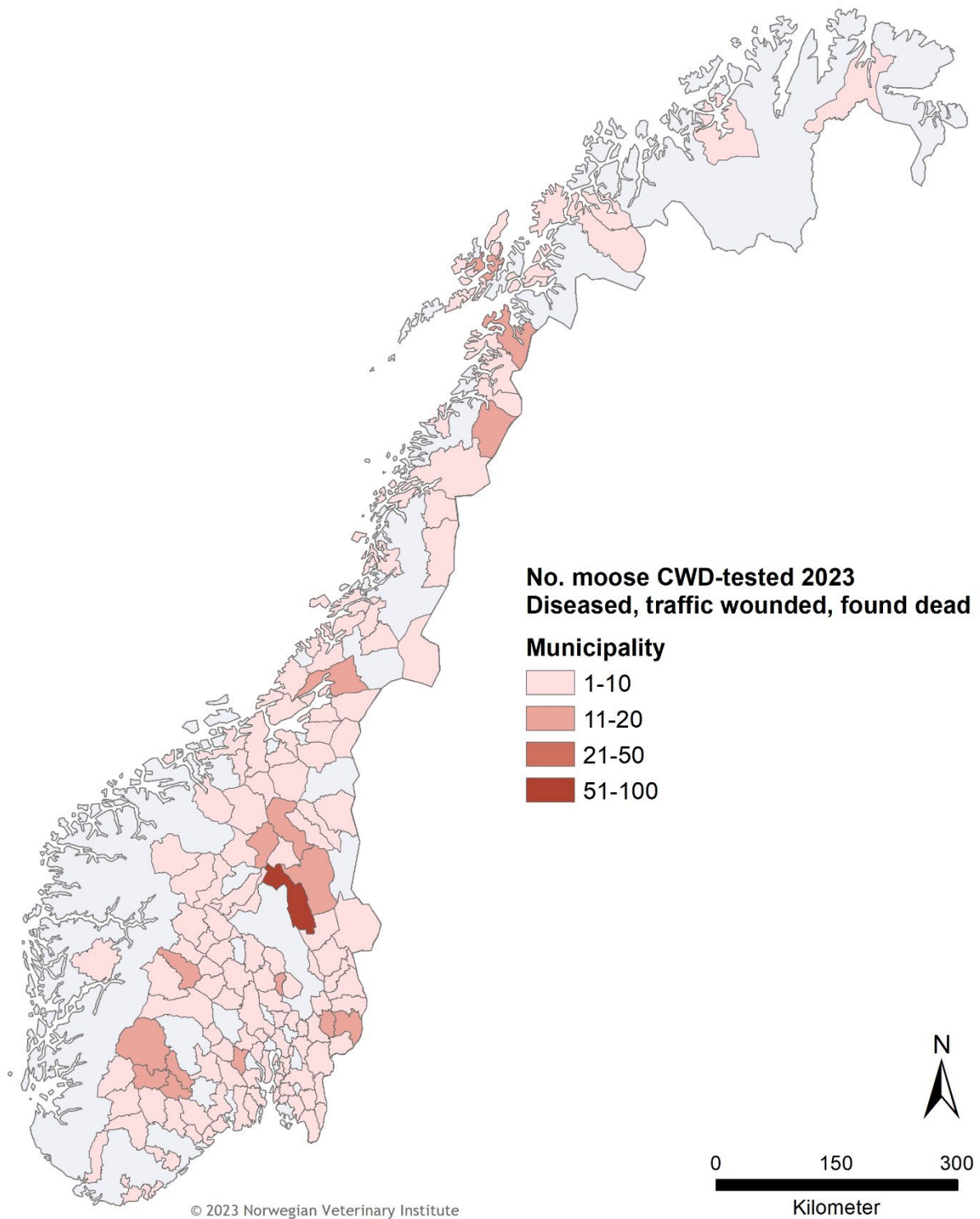


Figure 5: Number and geographical distribution of free-ranging moose (*Alces alces*) found diseased and euthanised, traffic wounded or dead and tested in the Norwegian surveillance programme for CWD in 2023.

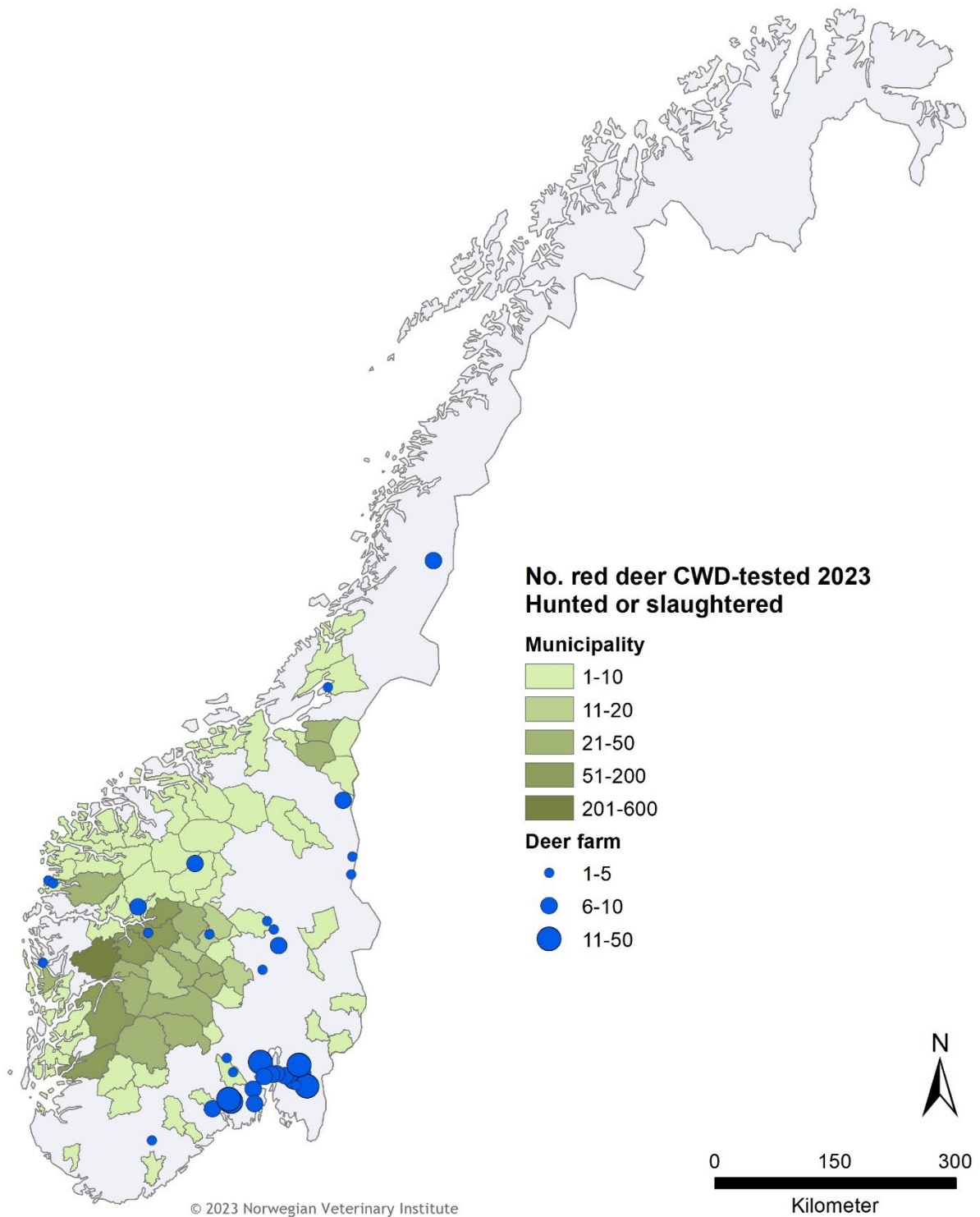


Figure 6: Number and geographical distribution of hunted free-ranging (green) and slaughtered captive (blue dots) red deer (*Cervus elaphus*) tested in the Norwegian surveillance programme for CWD in 2023.

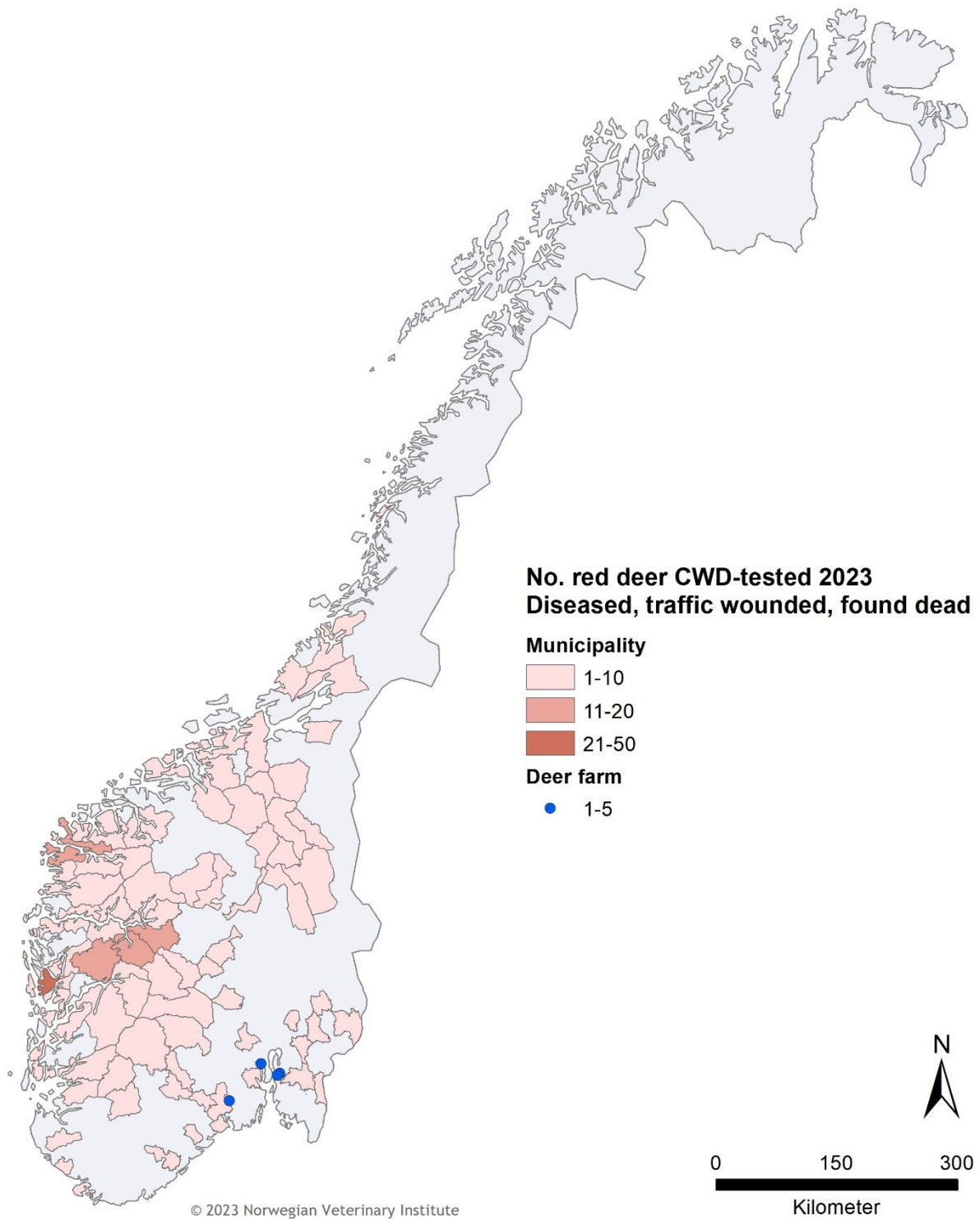


Figure 7: Number and geographical distribution of free-ranging (red) and captive (blue dots) red deer (*Cervus elaphus*) found diseased and euthanised, traffic wounded or dead and tested in the Norwegian surveillance programme for CWD in 2023.

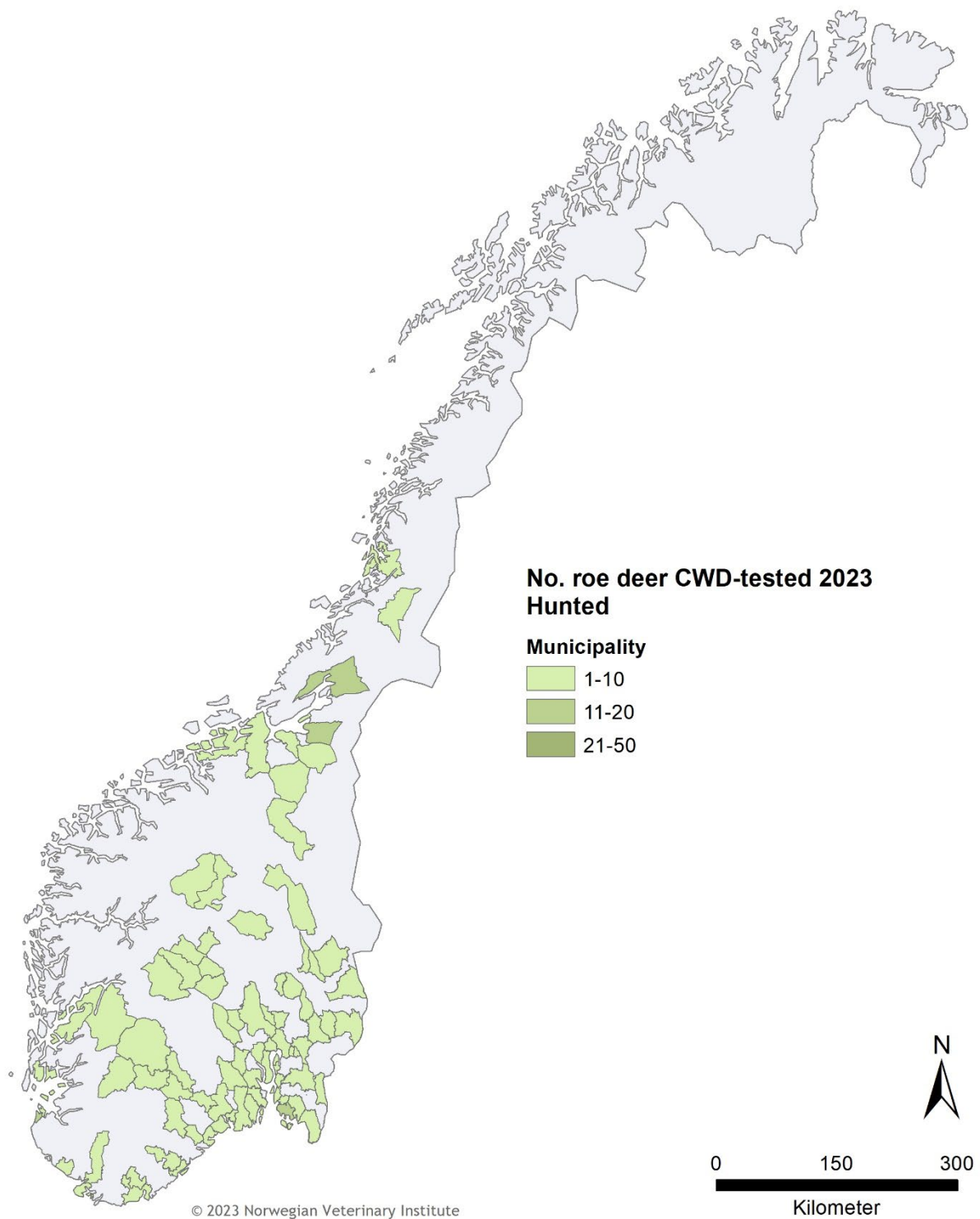


Figure 8: Number and geographical distribution of hunted free-ranging roe deer (*Capreolus capreolus*) tested in the Norwegian surveillance programme for CWD in 2023.

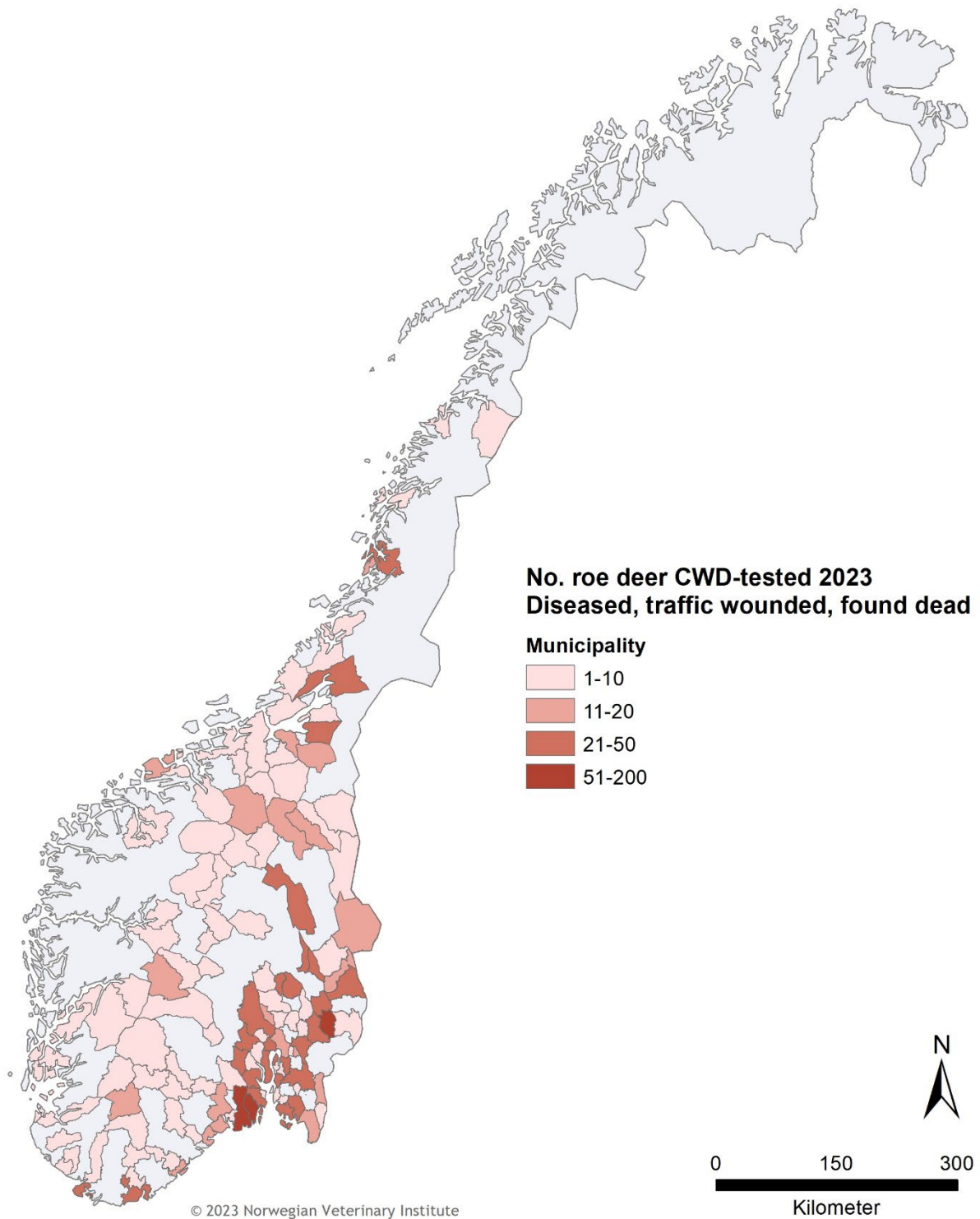


Figure 9: Number and geographical distribution of free-ranging roe deer (*Capreolus capreolus*) found diseased and euthanised, traffic wounded or dead and tested in the Norwegian surveillance programme for CWD in 2023.

Discussion

In 2016, CWD was for the first time detected in Europe, in six free-ranging cervids in Norway. In total, the CWD testing has revealed 21 reindeer, twelve moose and three red deer positive for the disease in the period 2016-2023. The first 19 reindeer cases were detected within the outbreak zone 1 of Nordfjella wild reindeer management area. The affected Nordfjella population was culled in 2018. Two additional cases have been diagnosed in reindeer hunted at Hardangervidda in September 2020 and September 2022. The discovery of CWD in a new reindeer area is challenging the prospect of eradicating the contagious disease in the reindeer populations.

Hardangervidda holds the largest sub-population of wild reindeer in Europe, and the area is much less confinable than Nordfjella in the aspect of disease spreading. Nevertheless, so far no additional reindeer cases have been detected outside Nordfjella and Hardangervidda management areas despite the testing of more than 86,000 reindeers. This indicates a limited infection in this species. Nevertheless, the probability of freedom as estimated using scenario tree modelling, varies a lot between different wild reindeer management areas or semi-domesticated reindeer districts depending on population size and number of tested animals per area or district. From the extended surveillance of cervids, we are confident CWD is not present at high prevalence (5%) in cervids in Norway [16]. For a lower prevalence (below 1%), we are less confident. Only a few wild reindeer management areas have obtained a 95 % probability of freedom for a design prevalence of 0.5 % [17]. The reindeer district of Filefjell, sharing border to the Nordfjella area, has obtained a high probability of freedom for a design prevalence less than 0.5 % [18].

Further research studies aiming at characterizing prion strain differences have shown that the strain found in reindeer is different from those of the two other cervid species. The reindeer cases were indistinguishable, in results of diagnostic test methods, from cases of CWD from North America. The positive European moose and red deer have shown atypical characteristics [6,7,14], contrasting with the reindeer and cervids from North America, with no detectable prions in the lymphoid tissues using diagnostic tests. Strains analysed in the Norwegian cases, across three species, were demonstrated by inoculation studies as multiple and all different from North American cases. Nevertheless, the reindeer strain has many similarities with strains isolated from cases from North America [19,20].

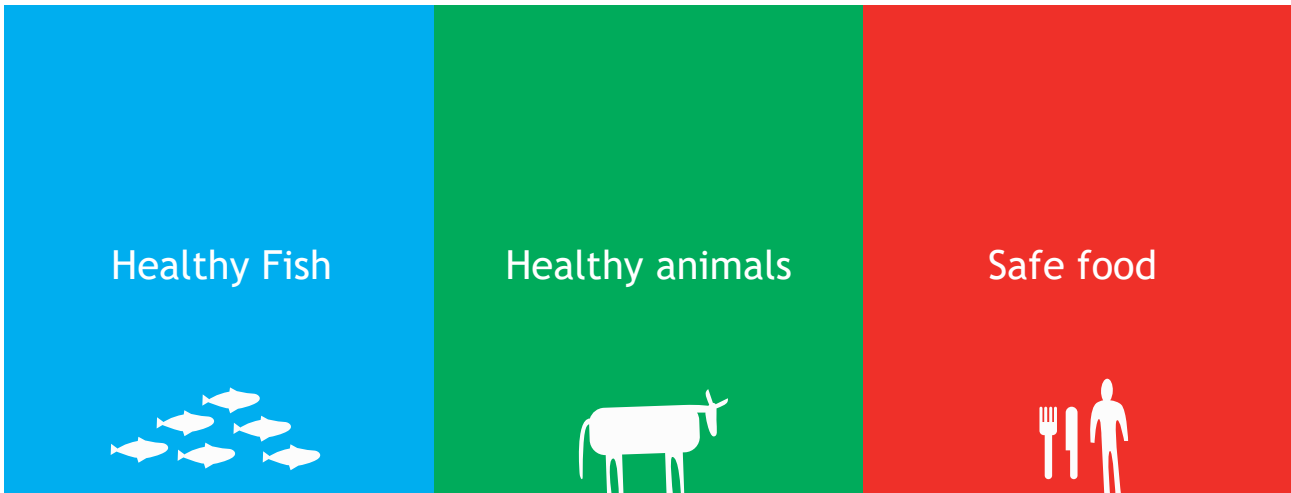
The positive CWD animals other than reindeer are 12 cases in 38,555 tested moose and three cases in 35,749 tested red deer, indicating low levels of horizontal spreading, if any, between live animals in these species [7]. TSE in domestic animals, with Nor98 / atypical scrapie in sheep and atypical BSE in cattle, have prion strains showing similar sporadic nature. Nor98 / atypical scrapie and atypical BSE have a prevalence of about eight per 10,000 found dead adult sheep and about two per 1,000,000 tested cattle, respectively.

The number of tested cervids in Norway is not enough to assure knowledge of accurate prevalence nor to give complete understanding of CWD epidemiology. For other European

countries there is even much more uncertainty as the surveillance levels are still very moderate [6].

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