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Factors that can contribute to spread of CWD – an update on the situation in Nordfjella, Norway

**Opinion of the Panel on biological hazards of the Norwegian Scientific Committee
for Food and Environment**

Report from the Norwegian Scientific Committee for Food and Environment (VKM) 2018: 16
Factors that can contribute to spread of CWD – an update on the situation in Nordfjella,
Norway

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Preparation of the opinion

The Norwegian Scientific Committee for Food and Environment (Vitenskapskomiteen for mat og miljø, VKM) appointed a project group to answer the request from the Norwegian Food Safety Authority. The project group consisted of one VKM member of the Panel on Microbial Ecology, two external experts and a project leader from the VKM secretariat. An external referee commented on and reviewed the manuscript. The VKM Panel on biological hazards evaluated and approved the final opinion drafted by the project group.

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Competence of VKM experts

Persons working for VKM, either as appointed members of the Committee or as external experts, do this by virtue of their scientific expertise, not as representatives for their employers or third party interests. The Civil Services Act instructions on legal competence apply for all work prepared by VKM.

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Summary

In spring 2018 the last wild reindeer in Zone 1 in Nordfjella wild reindeer area was shot. In total 2024 wild reindeer were killed, mostly through culling organized by the Norwegian Nature Inspectorate. The background for this measure, unparalleled in Norwegian game management, was the detection of the contagious and lethal Chronic Wasting Disease (CWD) in the spring 2016.

CWD is well known from North America where experiences and research have shown that disease is very difficult to eradicate. This is, among other things, due to infected animals going through a long incubation period when they shed infectious agents but do not show signs of disease. They act as silent carriers.

Infected animals can shed prions, for example through saliva, urine and faeces infecting other animals directly or by contaminating the environment where prions can then be picked up by a susceptible animal.

Another important aspect is that the infectious agent, misfolded proteins called prions, are very resilient. They can withstand radiation, heat and chemicals better than most contaminants we know. This allows them to "survive", ie, to be infectious in the environment for many years. The prions have also the ability to bind to soil, and they can be taken up in plants.

On that background, the Norwegian Food Safety Authority asked for an update of important factors affecting the spread of CWD within and out of the Nordfjella area.

CWD, in the form known from North America, has so far only been found in the wild reindeer population in Nordfjella Zone 1. Due to the resilience of prions, we must expect that they still persist in the areas used by the eradicated wild reindeer herd. Also, we cannot exclude that infected cervids can be found in the populations surrounding Nordfjella Zone 1. The prevalence of infected animals were low and the herd roamed over large mountain areas. In theory, this reduces the likelihood that new animals will contract infection from the environment. However, environmental conditions that promote aggregation of many animals in small areas will significantly increase the likelihood of contracting infection from the environment. A large number of mineral licks are placed in Nordfjella. These are attractive also for reindeer and other cervids. Many of the mineral licks are placed in areas used by the now exterminated wild reindeer population. Mineral licks and their surroundings, surfaces of both rocks, soil, plants, lichen could have been contaminated by saliva, urine and faeces from infected animals. As the soil around a mineral lick will be rich in salt, cervids and sheep will not only visit the lick itself but also lick or eat soil from the surrounding area. Since it is known that prions bind to and can survive for long time in soil, this could lead to prolonged exposure to infectious agents left behind by the extinct population.

Recent studies indicate that the prion isolated from the wild reindeer in Nordfjella is somewhat different from the strains found and investigated in North America. This may mean that the infectious potential and the development of disease in different species may deviate from what one would expect if it was similar prion type. However, we must assume that reindeer is easily susceptible to infection and has a disease development similar to that seen in North America. We must also assume that other cervids are susceptible, but we cannot tell how quickly the disease would develop. This, and the fact that cervids exposed to environmental contamination, may be exposed to low doses of contaminants, can cause the disease to develop slowly. In that case, the probability for silent carriers among cervids around Zone 1 would increase. Based on a combination of what we currently know about the degree of contact between populations and what we can say about susceptibility, we consider the probability for silent carriers within the Nordfjella area greatest for the wild reindeer population in Nordfjella Zone 2, the wild red deer in Lærdal and Aurland and the semi-domesticated reindeer in Filefjell. We assess the likelihood that there are such animals in the wild reindeer population in Lærdals-Årdalsfjella or red deer, moose or roe deer in other parts of the Nordfjella area to be lower.

Very high density of cervids in the areas around Zone 1 increases the likelihood of infection since the number of cervids visiting Zone 1 increases proportionally to population. We cannot say that mineral lick sites are important for attracting cervids into the area, but they represent points where the deer can still contract infection. A large population would also require more extensive testing to reach adequate level of certainty considering the absence of silent carriers. A significant reduction in cervid population would reduce the likelihood of new animals being infected and increase the likelihood of any contaminated animals being removed while increasing the knowledge of where the infection occurs.

Nordfjella has long been an important grazing area, not least for sheep. Every year there are several tens of thousands of sheep on summer pastures there. Most of the sheep belong in the area close to the Nordfjella, while others come from far away, on a so-called guest pasture. In recent decades, the use of man-made, permanent mineral licks has become common in sheep management. The licks help to supplement the sheep's salt needs and facilitate the observation and control of the animals on mountain pastures. However, in the areas previously occupied by the infected reindeer population, the mineral licks will increase the probability for the sheep to be exposed to CWD prions. We believe the likelihood that the sheep may become infected and develop disease is very small. However, a hypothetical and poorly investigated possibility is that the sheep can become silent carriers not developing the disease but still spreading small amounts of infectious agent. The probability of transmission of CWD from Nordfjella Zone 1 to other cervid populations by this mode is considered small, but cannot be excluded. In addition, sheep and other animals moving in Zone 1 can ingest prions which then are passed through the digestive tract, and wool and hooves can be contaminated with soil containing infectious material. However, a series of unfortunate circumstances must occur if CWD prions are to be transferred from Nordfjella to cervids in new locations in this way. This is very unlikely in a case of a single sheep, but as there are several tens of thousands of sheep on the pasture, this cannot be entirely disregarded. Sheep that has been on a guest pasture in Nordfjella, and then being transported home to a

completely different area and released on a farm there, represents a special challenge in this context, as the preparedness for and monitoring of possible outbreak of CWD supposedly is lower there than in Nordfjella area.

Other animals (scavengers, carnivores, rodents, etc.) inhabiting Nordfjella can also be considered to carry infection. The number of those animals, the degree of contact and/or the degree of susceptibility is assumed to be low, hence the likelihood of the transmission of contamination within or out of the Nordfjella area is considered to be very low. It has been shown that canids and crows ingesting prion-containing material are capable of passing infectious prions in their faeces, and this confer a low risk of spread.

The same applies also to mechanical transport of infection by humans via, for example, excrements and soil on boots, vehicles or equipment. It is not very likely that such transmission of infection would cause outbreak of CWD in other cervid populations. However, humans can move far and unpredictably, so the infection may appear in completely unexpected locations where there is no monitoring and preparedness.

More than 200 mineral licks are registered in the area where CWD was detected. In addition, there are another 500 mineral licks placed in other parts of the Nordfjella area. Mineral licks in the Zone 1 represent the greatest known potential source for further spread of CWD, both to nearby cervid populations and via sheep. The likelihood of spreading would be significantly reduced by removing these possible sources of infection.

Presence of the remaining mineral licks in the area and the placement of new ones would represent a hazard if infected animals still live in the area or if new animals get infected from the environment. Mineral lick sites would then most likely represent effective points for spread of infection. Reduction of the number of mineral licks would therefore be an important preventive measurement for reducing this probability of spread of infection.

An important issue in this context is that we do not yet have a simple, feasible method either for the detection of prions in the environment or for decontamination of mineral licks and surrounding areas. Therefore, the reduction of the numbers of those places would reduce the potential efforts related to a reoccurring outbreak.

Based on the current knowledge, but with considerable uncertainty, we have identified the following factors being able to contribute to the spread of CWD from and within the Nordfjella area:

1. Presence of undetected carriers of CWD in cervid populations around Zone 1; either animals that were infected before the wild reindeer population was eradicated or animals exposed to environmental contamination in Zone 1 after the eradication. The probability of this is considered to be greatest for the red deer in Lærdal and Aurland, the wild reindeer population in Zone 2 and semi-domesticated reindeer population in Filefjell.
2. Reindeer from Zone 2 or Filefjell that moves into Zone 1 and becomes infected.
3. Red deer, moose and roe deer from the nearby populations entering Zone 1 and becoming infected.

4. Sheep grazing in Zone 1 and carrying infectious material to susceptible cervid populations.
5. Human activity in Zone 1, transporting infectious material to susceptible cervid populations.
6. Other animals present in Zone 1 carrying infectious material from there to susceptible cervid populations.

In each of these cases, the number of currently available mineral lick sites previously used by eradicated wild reindeer population in Zone 1, have a great importance for the likelihood of spread of infection. The likelihood of cervids entering Zone 1 becoming infected would be greatly reduced if these sites were not available. Equally, the likelihood of sheep transferring the infection will be significantly reduced.

The likelihood that any carriers outside Zone 1 would spread the infection to new areas and that the disease becomes established there can be reduced by continued intensive monitoring, high alertness and preparedness. General reduction in the number of mineral lick sites and other places where cervids gather, reduction in the density of the nearby cervid populations, measures minimizing animal movements between areas and continued information to humans using the area would also help to reduce the likelihood of infection spreading.

Sammendrag på norsk

Våren 2018 ble den siste villreinen i Sone 1 i Nordfjella villreinområde skutt. Da var 2024 villrein avlivet, de fleste gjennom statlig felling organisert av Statens naturoppsyn. Bakgrunnen for dette tiltaket, som savner sidestykke i norsk viltforvaltning, var påvisning av den smittsomme og dødelige skrantesyken (Chronic Wasting Disease, CWD) våren 2016.

Sykdommen er godt kjent fra Nord-Amerika. Erfaringer og forskning derfra indikerer at skrantesyke er svært vanskelig å bekjempe. Dette har blant annet bakgrunn i at dyr som smittes går gjennom en lang inkubasjonsfase hvor de skiller ut smittestoff, men ikke viser tegn til sykdom. De fungerer som skjulte smittebærere.

Smittestoffet, feilfoldede proteiner kalt *prioner*, er svært hardført. Prionene tåler stråling, varme og kjemikalier bedre enn de fleste smittestoff vi kjenner. Dette gjør at de kan «overleve», dvs. holde seg smittsomme i miljøet, over mange år. Prionene har blant annet evne til å binde seg til jord, og de kan bli tatt opp i planter.

Smittede dyr kan skille ut prioner, for eksempel gjennom spytt, urin og avføring og slik smitte andre dyr direkte, eller ved at smitten blir værende i miljøet, for så å bli plukket opp av et mottakelig dyr.

Basert på dette ba Mattilsynet om en oppdatering av viktige faktorer som påvirker spredningen av skrantesyke (CWD) innenfor og ut av Nordfjellaområdet.

Smittsom skrantesyke, slik sykdommen er kjent fra Nord-Amerika, er til nå kun påvist i den nylig utryddede villreinbestanden i Sone 1. På grunn av smittestoffets hardførhet må vi regne med at smitten fortsatt finnes i områdene villreinen brukte. Vi kan heller ikke utelukke at smittede hjortedyr kan finnes i bestandene rundt Sone 1. Det ble funnet få smittede dyr spredt over store fjellområder. Det reduserer sannsynligheten for at nye dyr skal plukke opp smitte fra miljøet. Sannsynligheten for miljøsmitte vil imidlertid øke betydelig med faktorer som samler mange dyr på små arealer. I Nordfjella er det utplassert mange saltslikkesteiner. De er attraktive både for rein og andre hjortedyr. Mange av saltslikkesteinene har vært plassert i områder som den utryddede villreinbestanden har brukt. Overflater på både berg, jord, planter og lav, selve saltsteinen og stativet den står i, kan ha blitt forurensset med spytt, urin og avføring fra infiserte dyr. I og med at jorda rundt en saltslikkestein vil være rik på salt, vil hjortedyr og sau også slikke eller spise jord i området rundt. Siden det er kjent at prioner binder seg til og kan overleve lenge i jord, kan det medføre at dyra eksponeres for smittestoff fra den utryddede flokken i lang tid.

Nye undersøkelser indikerer at skrantesykeprionene fra villreinen i Nordfjella kan være forskjellige fra variantene som hittil er undersøkt i Nord-Amerika. Smitteevnen til og sykdomsforløpet kan derfor være annerledes enn det man ville ha forventet om prionstammene hadde vært helt like. Foreløpig funn indikerer at reinsdyr er mottakelige for smitte og har et sykdomsforløp som likner det som er sett i Nord-Amerika. Vi må også gå ut

fra at andre hjortedyr er mottakelige, men vi kan ikke si sikkert hvor fort sykdommen vil utvikle seg. Dette, og det at hjortedyr som blir utsatt for miljøsmitte gjerne blir eksponert for lave doser smittestoff, kan gjøre at sykdomsforløpet går sakte. Det vil i så fall øke sannsynligheten for at det kan finnes skjulte smittebærere i hjorteviltbestandene rundt Sone 1. Basert på en kombinasjon av det vi foreløpig vet om graden av kontakt mellom bestandene og det vi kan si om mottakelighet, mener vi sannsynligheten for skjulte smittebærere innenfor Nordfjellaområdet er størst for villreinbestanden i Nordfjella Sone 2, hjortebestanden i Lærdal og Aurland, og tamreinen i Filefjell. Vi regner sannsynligheten som mindre for at det finnes slike dyr i villreinbestanden i Lærdals- og Årdalsfjella eller i bestandene av hjort, elg og rådyr i andre deler av Nordfjellaområdet.

Svært høy tetthet av hjort i områdene rundt Sone 1 øker faren for smittespredning siden antallet hjort som besøker Sone 1 øker i takt med bestanden. Vi kan per dags dato ikke si at salteplassene er viktige for å trekke hjort inn i området, men de representerer punkter hvor hjorten fortsatt kan fange opp smitte. En stor bestand vil også kreve mer omfattende testing for å oppnå tilstrekkelig sikkerhet med tanke på skjulte smittebærere. En betydelig reduksjon av hjortebestanden vil være viktig for å redusere sannsynligheten for at nye dyr blir smittet, øke sannsynligheten for at eventuelle smittede dyr blir fjernet og samtidig styrke kunnskapen om hvor smitten finnes og ikke finnes.

Nordfjella har i lange tider vært en viktig beiteressurs, ikke minst for sau. Hvert år er flere titusener sauer på sommerbeite der. De fleste sauene hører hjemme i områdene rundt Nordfjella, mens andre kommer langveis fra, på såkalt gjestebeite. De siste tiårene har bruken av fastmonterte saltsteiner blitt vanlig i sauebruket. Disse bidrar til å dekke sauens saltbehov og letter tilsynet og kontrollen med dyrene på fjellbeite. Der hvor den smittede villreinflokket har vært, vil det imidlertid være fare for at sauen eksponeres for skrantesjukeprioner. Vi mener sannsynligheten for at sauen kan bli infisert og utvikle sykdom er svært liten. En hypotetisk og dårlig undersøkt mulighet er imidlertid at sauen kan bli en skjult smittebærer som ikke utvikler sykdom, men likevel sprer små mengder smitte. Sannsynligheten for at smitte skal spres på denne måten vurderes som liten, men kan ikke utelukkes. I tillegg kan sau og andre dyr som beveger seg i Sone 1, få i seg smittestoff gjennom munnen som deretter passerer gjennom fordøyelseskanalen, og ull og klauver kan bli tilskitnet med jord som inneholder smittestoff. Om skrantesjukeprioner skal overføres fra Nordfjella til hjortedyr i nye områder på denne måten, må det skje et sammentreff av en rekke uheldige omstendigheter. Det er svært lite sannsynlig i det enkelte tilfelle, men i og med at det er flere titusen sauer på beite, kan man ikke se helt bort i fra at dette kan skje. Sau som har vært på gjestebeite i Nordfjella, men deretter transporteres hjem til et helt annet område og slippes på beite der, representerer i denne sammenhengen en særlig utfordring, da beredskapen, overvåkingen og årvåkenheten ovenfor et eventuelt utbrudd av skrantesjuke kan være lavere der enn i Nordfjellaområdet.

Andre dyr (åtselere, rovdyr, gnagere mv.) som beveger seg i Nordfjella, kan også tenkes å ta med seg smitte. Antallet dyr, graden av kontakt og graden av mottakelighet antas å være

lav, og sannsynligheten for at transport av smitte med disse skal forårsake spredning innen eller ut av Nordfjellaområdet, må regnes for å være svært liten.

Det samme gjelder for mekanisk transport av smitte med mennesker, f.eks. møkk og jord på støvler, kjøretøy eller utstyr. Det er lite sannsynlig at en slik transport av smitte vil forårsake utbrudd i nye bestander av hjortedyr. Det vil imidlertid være en utfordring at mennesker kan bevege seg langt og uforutsigbart, slik at smitten kan dukke opp i helt uventede områder hvor det ikke er overvåking, årvåkenhet og beredskap.

I området hvor skrantesyke ble påvist er det registrert mere enn 200 saltsteiner. I tillegg finnes det ytterligere 500 salteplasser i andre deler av Nordfjellaområdet. Salteplassene i Sone 1 representerer den største kjente, potensielle kilden til videre spredning av skrantesjuka både til nærliggende hjorteviltbestander og med sau. Sannsynligheten for spredning av skrantesyke vil reduseres betydelig ved fjerning av disse mulige smittekildene.

Opprettholdelse av de resterende saltsteinplassene i området og opprettelse av nye, vil representere en fare dersom det skulle finnes infiserte dyr i området eller om nye dyr skulle bli smittet fra miljøet. Saltsteinplassene vil da med stor sannsynlighet fungere som effektive stasjoner for smittespredning. Å redusere antallet saltsteinplasser vil derfor være viktig for å forebygge smittespredning.

Et moment i denne sammenhengen er at vi ennå ikke har en enkel gjennomførbar metode verken for sikker påvisning av smitte eller for sanering av smitte på saltsteinplasser. Dersom antallet saltsteinplasser ble redusert, vil innsats og kostnad knyttet til eventuell sanering og senere opprydning reduseres.

Vi har på grunnlag av det vi i øyeblikket vet, men med betydelig usikkerhet, identifisert følgende forhold som kan bidra til spredning av skrantesjuka ut fra og innenfor Nordfjellaområdet:

1. Forekomst av uoppdagede smittebærere blant hjortedyr i bestandene rundt Sone 1: Enten dyr som var smittet før villreinen ble utryddet, eller dyr som har blitt eksponert for miljøsmitte i Sone 1 etter utryddelsen. Sannsynligheten for dette vurderes som størst for hjorten i Lærdal og Aurland, villreinen i Sone 2 og tamreinen i Filefjell.
2. Rein fra Sone 2 eller Filefjell går inn i Sone 1 og blir infisert.
3. Hjort, elg og rådyr fra bestandene rundt går inn i Sone 1 og blir infisert.
4. Sau som beiter i Sone 1 og bærer med seg smitte ut derfra til mottakelige hjortedyrbestander.
5. Menneskelig aktivitet i Sone 1 som medfører at smitte blir transportert ut derfra og til mottakelige hjortedyrbestander.
6. Andre dyr som beveger seg i Sone 1 og tar med seg smitte derfra og til mottakelige hjortedyrbestander.

I hvert av disse forholdene vil antallet tilgjengelige salteplasser som villreinbestanden i Sone 1 har brukt, ha svært stor betydning for sannsynligheten for smittespredning. Om disse salteplassene ikke var tilgjengelige, ville sannsynligheten for at hjortevilt som går inn i Sone 1 blir smittet, bli svært mye mindre. Likeledes vil sannsynligheten for at sau tar med seg smitte bli betydelig redusert.

Sannsynligheten for at eventuelle smittebærere utenfor Sone 1 sprer smitten videre til nye områder og at sykdommen skal etableres der, kan minimaliseres med fortsatt intensiv overvåking, stor årvåkenhet og slagkraftig beredskap. Generell reduksjon av antallet salteplasser og andre samlingssteder for hjortevilt, bestandsreduksjon i kontaktbestandene, tiltak som minimaliserer dyras bevegelse mellom områder og fortsatt informasjon til mennesker som bruker området, vil også være med på å redusere sannsynligheten for smittespredning.

Background and terms of reference as provided by the Norwegian Food Safety Authority

The Norwegian Food Safety Authority has requested an update and possibly elaboration on important factors influencing the spreading of Chronic Wasting Disease (CWD) within and out of the Nordfjella-area. We refer to the risk assessments on CWD (phase 1 and 2) and the statements dated 29.9.2017 and 2.2.2018 updating the assessments.

The assessments were important basis for the preparation of the Plan for the Eradication of the Wild Reindeer Population in Nordfjella Zone 1 and the Plan for the reestablishment of the Reindeer Population in Nordfjella Zone 1. Many of the factors are already discussed in the VKM-opinions, while others are less, or not, discussed.

The last wild reindeer in Zone 1 was killed in the spring (2018) and the eradication process is now in the following phase. To succeed with the following and the reestablishment of wild reindeers in Zone 1, it is important that factors that can contribute to the spreading of the infectious agent within and out of the Nordfjella-area are limited.

Further, the situation has changed since the assessment reports were written. In addition to the wild reindeers in Zone 1 being decimated, all saltlick in the Nordfjella has been completely or partly fenced off and substituted by new, open saltlicks. There is an ongoing process to reduce the population of cervids in the border zones of Nordfjella. A total of 45 000 samples have also been analysed since the first case in the spring 2016. Other measures are regulated in the Regulation on CWD Zone and the Regulation on CWD.

Based on this, we request an update and possibly elaboration on important factors influencing the spreading of Chronic Wasting Disease (CWD) within and out of the Nordfjella-area.

1 Introduction

March 2016, a reindeer (*Rangifer tarandus tarandus*) in Nordfjella Wild Reindeer Area in Norway, was diagnosed with Chronic Wasting Disease (CWD). This was the first observation of CWD in Europe, and the first natural case of prion disease in reindeer (REF).

Since then, 18 more reindeer cases were diagnosed in the same area, confirming that this was an outbreak of an infectious disease. In addition, four Norwegian cases of CWD have been diagnosed in moose (*Alces alces*) and one in red deer (*Cervus elaphus*) in areas relatively distant from Nordfjella and each other.

The Norwegian Food Safety Authority (NFSA) consulted the Norwegian Committee for Food Safety and Environment (VKM), for scientific opinions concerning zoonotic potential, transmission between animals and other relevant aspects, such as prion diseases and strain variability. The Phase I report, released in June 2016, assessed the situation primarily with reference to experiences with CWD management in the United States and Canada.

The Phase II report (VKM, 2017b), gave an update and a detailed discussion of managerial options, including that of eradication. Based on these opinions and after joint consultations with the Norwegian Veterinary Institute, the Norwegian Environment Agency and Norwegian Institute for Nature Research, NFSA concluded that measurements outlined by VKM, including the eradication of the wild reindeer population in Nordfjella Zone 1, should be implemented rapidly. The Norwegian Ministry for Food and Agriculture approved this and ordered a Management and Repopulation Plan (Miljødirektoratet & Mattilsynet, 2017). This plan should include practical descriptions on how the affected herd should be eradicated within the 1st of May 2018 including animal welfare aspects, and how a population could be reestablished in the affected area after a fallow period.

In addition, the NFSA requested two further statements from VKM, both addressing the zoonotic potential of CWD in view of data from experimental transmission of CWD to macaques and the emergence of what appears to be new CWD strains in Norway. These statements were issued in 2017 (VKM, 2017a) and 2018 (VKM, 2018) and concluded that the zoonotic risk of CWD given as “very low” in the Phase I report should be maintained.

Most of what is currently known about CWD is based on research and experiences from North America and prion research in general. In addition, we can extrapolate some knowledge from research on and experiences with other transmissible spongiform encephalopathies in animals and humans, most notably scrapie in sheep, bovine spongiform encephalopathy (mad cow disease) in cattle and Creutzfeldt-Jacobs disease in humans. However, the uncertainties regarding the validity of such comparisons must be kept in mind. For instance, there might be important differences between the Nordfjella prion isolate and those known from North America.

Based on North-American assessments, there are three properties of CWD that signal that strong measures are warranted to combat the spread of disease:

1. CWD seems to have a major population impact on at least some of the affected populations and can, according to models, have the potential to eradicate a cervid species locally.
2. The disease has a very long course and it can be anticipated that the affected individuals and their family group experience severe stress and suffering during a considerable part of that period.
3. The zoonotic potential of CWD is regarded to be very low, but not negligible. To minimize the probability of spill-over from cervids to humans (and other species), it is important to minimize distribution and prevalence of the disease among cervids.

This report provides a knowledge background for Norwegian authorities concerning factors that can contribute to the spread of Chronic Wasting Disease (CWD) within and out of the Nordfjella-area and by this indicate measures that can be considered in order to prevent spread and increase the likelihood of successful eradication of this disease from Norway.

1.1 Definitions of areas described in this report

A regulation establishing preventive measures against potential spread of CWD from Nordfjella was put in action on the 14th of June 2017 ((LMD, 2017)). This regulation defined the Nordfjella Management Area as the complete area of the municipalities Ulvik, Lærdal, Aurland and Hemsedal and Eidfjord, Hol and Ål north of the road RV7. Recent revisions of the regulation excluded the areas of Ulvik municipality west of the river Tysso from the Management Area (Mattilsynet, 2018). Consequently, the Nordfjella Management Area covers an area much larger than the Nordfjella wild reindeer area. In addition, the wild reindeer area is divided by a road (FV50 Hol – Aurland) into two zones called Zone 1 (north of the road) and Zone 2 (south of the road). Consequently, we speak about five different geographical entities (Figure 1-1):

1. Nordfjella wild reindeer area (NF-WRA) is a management area for wild reindeer. Wild reindeer areas are defined as the area above the forest limit used by a certain wild reindeer population (according to the Wild Cervids Management Regulation, Forskrift om forvaltning av hjortevilt). In some areas, where reindeer also use forested areas, these can be included, but this is not the case in Nordfjella. NF-WRA is divided in two zones (Zones 1 and 2).
2. Nordfjella Zone 1 (NF-Z1) is the northern part of the wild reindeer area, delimited by the highway E16 in the north, and the roads RV52 in northeast/east and RV7/FV50 in the south, and the fjord Sognefjorden in the west.
3. Nordfjella Zone 2 (NF-Z2) is the southern part of the wild reindeer area, separated from the larger Zone 1 by the road FV50 crossing the mountain area. To the west the area is delineated by steep cliffs and the railroad Flåmsbana. To the south the railroad Bergensbanen was regarded as an obstacle for southwards movement of the reindeer,

but this was relieved by opening of the 10.3 km railroad tunnel Finsetunnelen in 1993, and reindeer from the Nordfjella population now cross the tunnel roof over to the northern part of Hardangervidda reindeer area (see below).

4. Nordfjella Management Area (NF-MA) is the chronic wasting disease management area established by the authorities according to regulation as described above, delineated by the administrative borders of the municipalities (Figure 1-1), the highway RV7 in the south and the river Tysso in the southwest.
5. Nordfjella Zone 0 (NF-Z0) is the area of Nordfjella Management Area which not is included in the wild reindeer area (Zones 1 and 2).

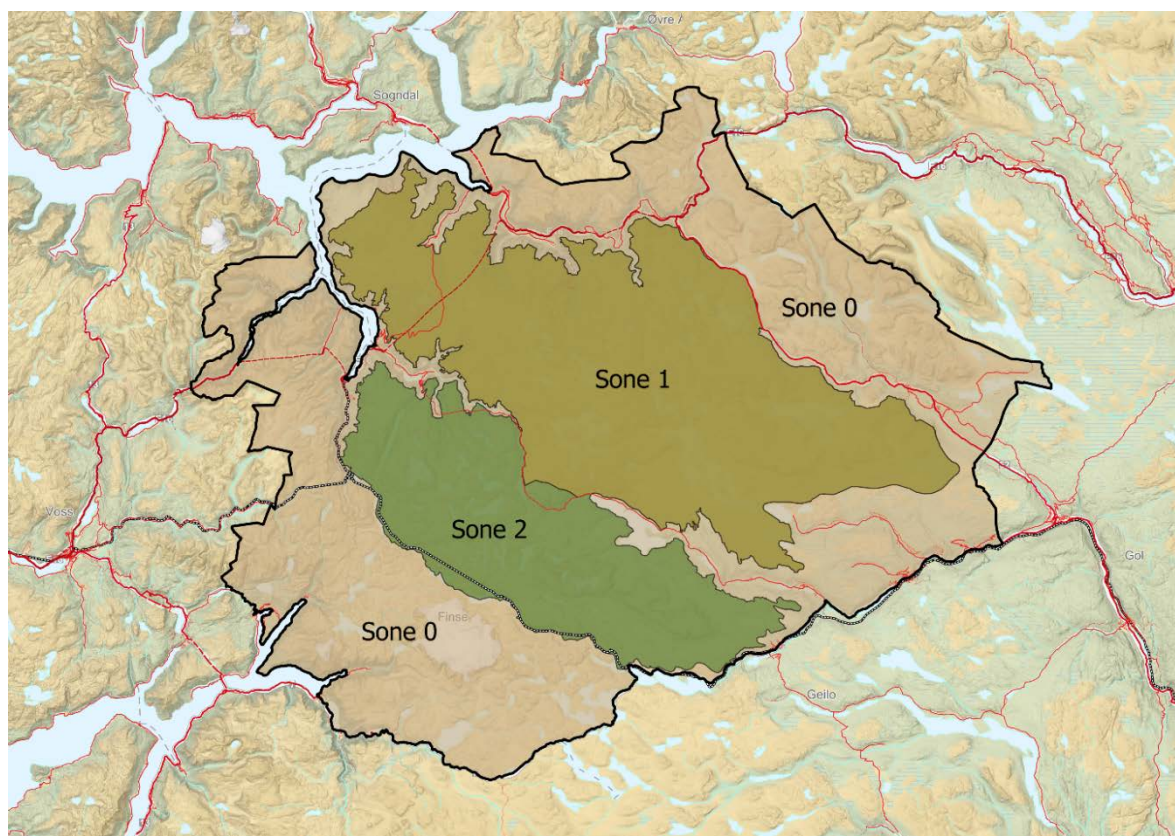


Figure 1-1. The Nordfjella Management Area (NF-MA) consists of the complete area of the municipalities Aurland, Lærdal and Hemsedal, the municipalities of Ål, Hol and Eidfjord north of the highway RV7 and Ulvik municipality east of the river Tysso. Within this area, Nordfjella wild reindeer area (NF-WRA) covers a large proportion of the most alpine parts but is divided in a northern Zone 1 (NF-Z1) and a southern Zone 2 (NF-Z2) by a crossing road (FV50).

1.2 Management actions already enforced

As described by the Ministry for Food and Agriculture, NFSA and the Norwegian Environment Agency developed a management plan for eradication of the wild reindeer herd in Zone 1. To provide a foundation for this work, several working groups were established to provide practical advice on how to implement the recommended measures. An overview of this work and resulting recommendations are given in "Saneringsplan for uttak av villreinbestanden i

Nordfjella Sone 1" (Plan for eradication of the wild reindeer population in Nordfjella Zone 1) and "Reetableringsplan for villreinbestanden i Nordfjella Sone 1" (Plan for repopulation of Nordfjella Zone 1).

1.2.1 Eradication of affected herd

For a thorough discussion of management options and scenarios, please see the Phase II report from VKM (VKM, 2017b). The conclusion was that for a semi-isolated reindeer population, the highest probability of eradication of CWD and least probability of disease dissemination would be achieved by eradicating the affected herd.

In accordance with this, Norwegian authorities decided to eradicate the wild reindeer population in NF-Z1 (Miljødirektoratet & Mattilsynet, 2017). This was accomplished first by extending the regular hunting season (582 animals) and thereafter by teams of sharpshooters organized by the Norwegian Nature Inspectorate. The last reindeer in Zone 1 was shot on the 11th of April, making the total number of animals culled 2024. In addition, 23 animals killed by avalanches during the hunting season were sampled.



Figure 1-2. Photos taken by NFSA during culling of the reindeer population in NF-Z1. All animals were tested for CWD and thereafter destructed.

1.2.2 Nation-wide surveillance for CWD

An unrecognized outbreak of CWD in a cervid species would increase the probability of spread even more than verified cases, since few precautions and counteractive measurements would be put efficiently in function, even in a situation where a known outbreak occurs within the same country (as in the case with Nordfjella). In addition, knowledge about the distribution of CWD was critical for the choice of adequate actions and was necessary to argue against the common perception that CWD had been present in Norway for ages, was widely distributed in cervid populations all over the country and any action to control or eradicate the disease consequently doomed to be unsuccessful. As shown in the table below (Table 1-1), a massive surveillance effort has been carried out since 2016, reaching a total of more than 64 000 cervids tested, by mid November 2018.

Table 1-1. Overview of cervids tested for CWD in Norway since 2004.

Species	2004-2015	2016	2017	2018*	Sum	CWD+
Moose (<i>Alces alces</i>)	130	4 403	5 468	6 402	16 403	4
Reindeer (<i>Rangifer tarandus tarandus</i>)						
Semi-domesticated	966	1 739	10 934	7 366	21 005	0
Wild (free-ranging)	10	838	2 912	3 673	7 433	19
Red deer (<i>Cervus elaphus</i>)	820				820	
Farmed		129	444	539	1 112	0
Wild (free-ranging)		2 453	3 637	6 568	12 658	1
Roe deer (<i>Capreolus capreolus</i>)	183	484	1 959	1 915	4 541	0
Fallow deer (<i>Dama dama</i>)	12	15	20	17	64	0
Total	2 121	10 061	25 374	19 470	64 036	24

In the NF-MA, 900 semi-domesticated reindeer, all from Filefjell tamreinlag (Filefjell reindeer breeders' co-operative), north of NF-Z1 have been tested and in NF-Z2 approximately 150 wild reindeer have been analysed, all with negative results. In addition, a total of 4094 wild red deer, moose and roe deer, all from the NF-MA have been tested and found negative. These data are clearly encouraging but continued enhanced surveillance is needed to document the absence of the disease. For instance, in NF-Z2, the sample size of 150 animals is insufficient to estimate a disease prevalence with adequate certainty. Based on the current number of tested animals, the Norwegian Veterinary Institute has by the use of a published model (Viljugrein et al., 2018) estimated the probability of absence of CWD to be around 64% for NF-Z2 and 67% for the Hardangervidda area.

In addition to surveillance in the NF-MA, testing of all cervids slaughtered, hunted or found dead, have been performed in more than 50 municipalities across Norway.

1.2.3 Efforts to eliminate “hotspots” for transmission

VKM phase II identified “elimination of hotspots for potential disease transmission in areas where contagious CWD has been diagnosed and in nearby areas where spread from initial cases is plausible” as important for reducing the risk of transmission. This is in accordance with North-American management and contingency plans for CWD, such as the quotation below from the recently published “Recommendations for Adaptive Management of Chronic Wasting Disease in the West” (WAFWA, 2017):

“Reduce Artificial Points of Host Concentration: Identify consistently available, artificial point-sources of food/minerals/water causing deer to aggregate (e.g., leaky grain bins, grain bags, stack yards, artificial feeders or feeding stations, mineral bins). Work with producers, landowners, and agriculture authorities to mitigate the point source and reduce the density of deer at these point-sources.”

As described above, a working group provided background for the practical management of mineral licks in NF-MA. The Zone Regulation of 12th of June 2017 described that all mineral licks within NF-MA should be made inaccessible for cervids. The compromise the working group reached, was that mineral licks considered to constitute a very high probability of transmission from reindeer to red deer, should be fenced off completely. Likewise, mineral licks established on soil (in contrast to gravel or bare rock) were considered to be high risk hot spots. Mineral licks not considered to constitute a major probability for transmission to red deer, should also be fenced off, but the fences could be equipped with openings that should allow access for sheep during the grazing period, while cervids were kept out. Where a mineral lick had been completely closed off, but the need for a mineral lick was regarded to be necessary, the sheep herders were allowed to establish a new one in the vicinity. These should be located on bare rock or well-draining sand/gravel. During the winter, the openings in the semi-closed exclosures were supposed to be closed and the mineral stones themselves be removed. The fence modules were 200 cm long and 184 cm high. The spacings between the vertical balusters were 15 cm wide, and the openings intended for sheep were 40 cm wide and 53,5 cm high (Figure 1-3). Where a mineral lick was closed off completely, ten modules without openings were connected to each other with plastic strips. Where an existing mineral lick should be accessible for sheep, eight modules, three with openings and five without, were connected. Where a new mineral lick was established, six modules, two with openings and four without, were connected. Mounting of the fence modules started in the first part of July 2017, but already the 17th of July two lambs were observed stuck with their hips between the balustrades (Figure 1-4). The process was now halted, and production of new fences with less wide (7 cm) spacings between the balustrades initiated. In addition, existing fences were enforced with a wire mesh on the lower half of the fence.

NFSA identified, by the help from sheep farmers, 684 man-made mineral licks within NF-MA (Figure 1-7). 278 of these were within NF-Z1, 143 in NF-Z2 and 263 in NF-Z0. In the end of August 2017, only 10 – 15 mineral licks were fenced off. The work was, however, continued, and in March 2018 60% of the spots in NF-Z1 were found to be closed. This improved to 70% in April. At present, 644 (268 (96.4%), 138 (96.5%) and 238 (90.5%)), respectively) of the mineral licks are closed down (numbers provided by NFSA). Based on information from landowners and sheep herders, 170 mineral licks (60%) in NF-Z1, believed to be visited by reindeer, red deer or moose (in addition to sheep) were closed completely off. In addition, 6 mineral licks in Filefjell Mountains were closed to prevent semi-domesticated reindeer from getting stuck inside them. Of the remaining 508 mineral licks, 464 were equipped with fences with sheep openings. The 40 remaining were not fenced due to various reasons; some were not found by NFSA, some had been covered with gravel so that fencing was not necessary and some were just left without fences. 18 of these 40, which are inside Zone 1, are according to NFSA decided to be completely closed. Where a mineral lick has been completely closed, the sheep herders have been allowed to establish a new mineral lick without fences at least 30 m from the original one. It was intended that salt blocks on both these and on the semi-closed ones would be removed at the end of the grazing season for sheep, and it was also intended that the openings in the semi-closed ones would be closed

by a purpose-made lid. However, inspection of semi-closed mineral licks in November 2018 revealed that the lids not had been put in place in 38 of 98 fences and that several salt stones still were left on some of the licks (source: NFSA).

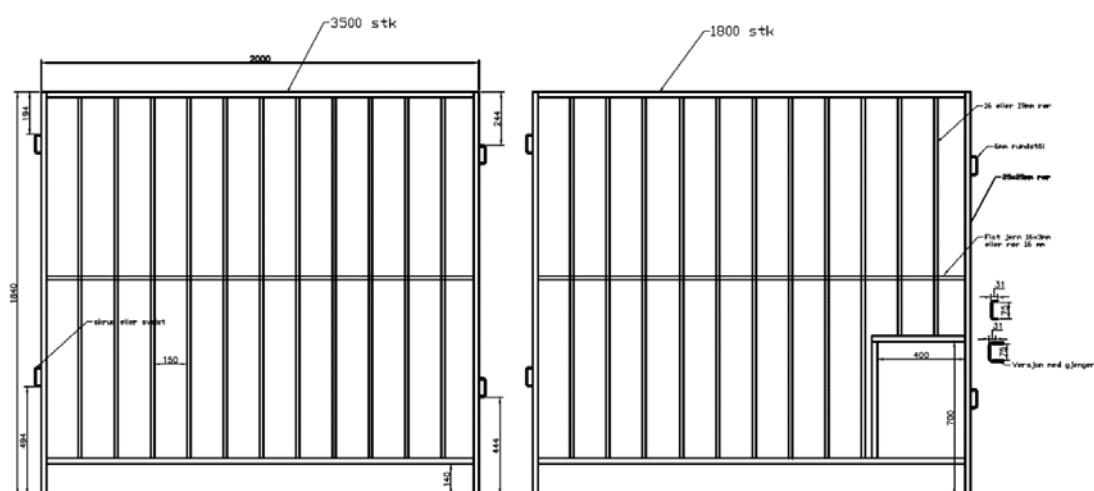


Figure 1-3. Original drawing of the fence modules without (to the left) and with (to the right) opening intended to allow sheep entrance to the mineral licks while cervids were kept out. (Drawing provided by NFSA).



Figure 1-4. Two lambs that were found trapped between the balustrades in the fences, caused a major halt in the process of fencing of the mineral licks in the summer of 2017. (Photo provided by NFSA).



Figure 1-5. A mineral lick enclosure in NF-Z2 is virtually filled with wild reindeer that easily go in and out through the openings. (Photo: NINA Wildlife Camera)



Figure 1-6. Red deer visiting a mineral lick enclosure in NF-Z1 together with an ewe and her lambs. (Photo: NINA Wildlife Camera)

Already in August 2017, a reindeer calf was observed inside exclosures with openings for sheep. In September 2017, also red deer were observed to enter the exclosures through the sheep openings. Footage from wildlife cameras later documented that both red deer and reindeer seemed to get habituated to the fences and learned to go in and out (Figure 1-5 and Figure 1-6). The openings consequently allow both red deer and reindeer access to the mineral licks.

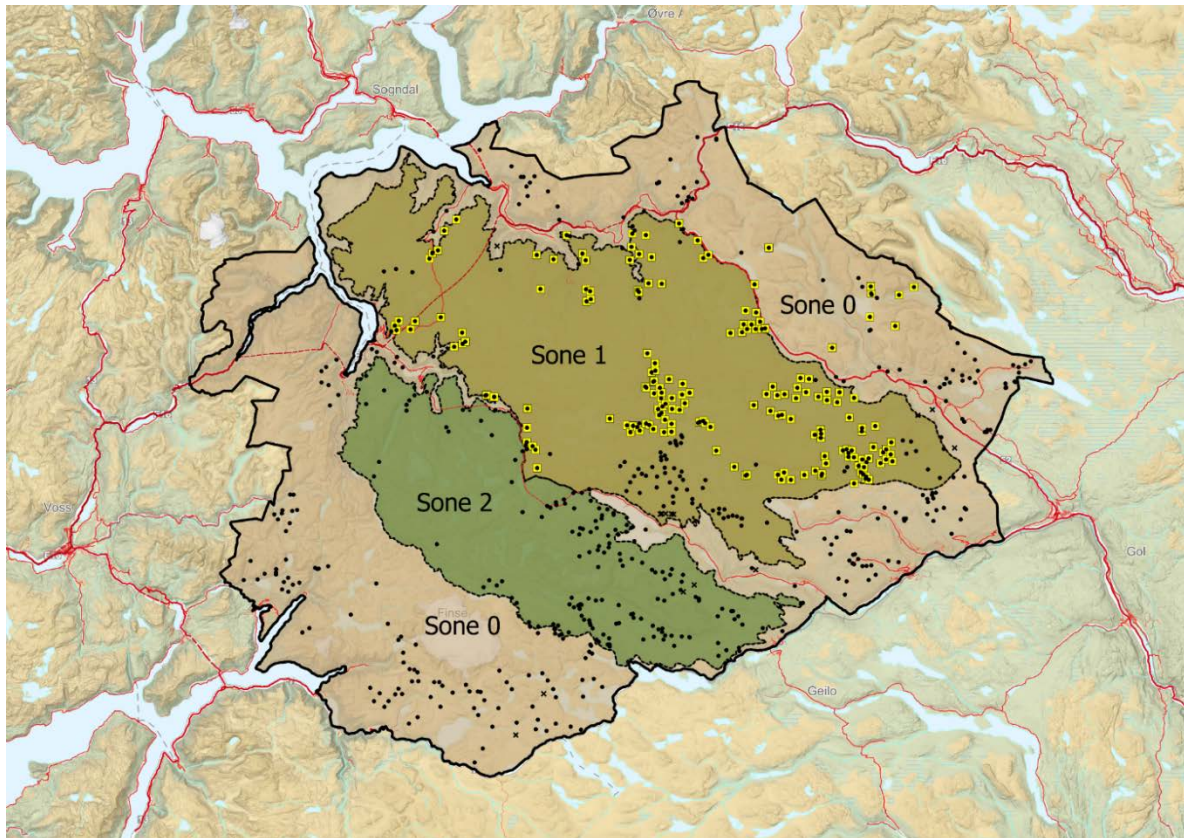


Figure 1-7. Permanent supplementary mineral licks for sheep identified in NF-MA. The 176 marked with a black dot within a yellow square have been fenced off, while 464 of the remaining 508 have been closed with fences with openings that supposedly should have allowed sheep access, while cervids were kept out. The crosses mark 13 mineral licks that not have been in use for over 5 years, are covered by at least 30 cm gravel or never have been used (although registered).

1.2.4 Efforts to minimize entry and transit of wild or semi-domesticated reindeer into Zone 1

In the winter of 2017, herders from Filefjell tamreinlag (Filefjell Reindeer Breeders' Co-operative) were allowed to chase back wild reindeer that approached the highway RV52. After a while, they were also allowed to shoot wild reindeer they not managed to stop. At the same time, personnel from the Norwegian Nature Inspectorate and Aurland fjellstyre (Aurland Mountain Management Board) followed the movements of wild reindeer along the roads RV7 and FV50, both by tracking/visual inspection from the roads and by following the positions of GPS-collared animals. If reindeer approached the roads, they were chased back.

If necessary, the personnel were allowed to shoot individual animals in order to stop and change the direction of the herd.

To minimize contact between wild reindeer in NF-Z1 and semi-domesticated reindeer in Filefjell, 145-170 cm high reindeer fences were built on stretches where the animals were known to cross RV 52. The fences, which were placed on the southern side of the road, are 14 km long in total. In the summer of 2018, however, semi-domesticated reindeer crossed the road in new places, and additional 11 km of new fences were built. In addition, 2 km of sheep fence were made higher. The semi-domesticated reindeer is herded continuously during the season when they graze on Filefjell, and the herders patrol along the road (and fences) several times a day in this period.

Fences has also been put up along FV50 to hinder wild reindeer from NF-Z2 from moving northwards. These fences are 160 cm high and 9,2 km long (pers. comm. A. Mysterud). In periods of deep snow, the fences are temporarily reinforced with sticks and bands, providing visual barriers for the reindeer.

1.2.5 Population reduction in neighbour areas

In April 2017, the Environmental Agency sent a letter to the municipalities in the NF-MA and the neighbouring municipalities of Årdal, Vang, Vestre Slidre, Nord-Aurdal, Sør-Aurdal, Gol, Nes, Nore og Uvdal, Voss and Vik in which they strongly recommended an increase of the hunting quotas for red deer, moose and roe deer. The agency did not specify any proportional aim for the increase but suggested that the quotas could be increased with 20% or that one could increase hunting on adult females. In 2017 the hunting bag of moose in the area (not including Voss and Vik) increased from 1239 to 1278 (3%) compared with the average in 2014 – 2016, while number of shot red deer increased from 1114 to 1804 (62%) and number of shot roe deer increased from an average of 99.5 to 104 (5%). In Lærdal, Aurland and Årdal together, 1196 red deer were shot in 2017, compared with an average of 670 the three previous years. Among the important success factors for this accomplishment was provision of free helicopter transport of carcasses in Aurland and Lærdal and the use of a game handling establishments that could ease the handling of the meat. In addition to increased hunting, reports from wildlife managers in NF-MA indicate that an additional population reduction occurred associated with winter starvation due to long-lasting and deep snow cover and late arrival of spring.

1.3 Scientific projects initiated

1.3.1 Bioassay experiments in laboratory rodents

Experimental transmission of primary prion isolates to laboratory rodents is important for analysis of prion strain properties and comparison of isolates. Such experiments are currently ongoing in which material from Norwegian CWD cases is used for inoculation (coordinated by

the Norwegian Veterinary Institute). Publication of results from inoculation in transgenic mice and bank voles (*Myodes glareolus*) is expected by mid-2019 (S. L. Benestad, personal communication). We are aware of preliminary results that support the notion of significant strain variation among Norwegian CWD isolates. However, a broader discussion of this must await the published reports.

1.3.2 Oral inoculation of newborn lambs with VRQ/VRQ PrP genotype

During spring 2018, a group of newborn lambs were inoculated orally with infectious material from a verified CWD case from Nordfjella. A schematic overview of these experiments is given in Figure 1-8. This experiment, performed in NMBU's research facility in Sandnes, is currently ongoing.

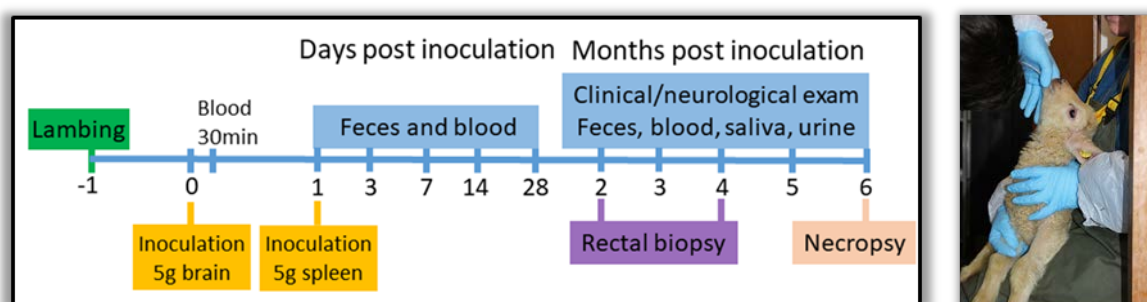


Figure 1-8. Experimental plan for oral inoculation of newborn lambs with brain and spleen material from a verified CWD case from Nordfjella. Animals have not been sacrificed as indicated, since the study has been extended (Photos: Cecilie Ersdal, NMBU).

Rectal biopsies have been collected from the inoculated animals and will be processed for analysis in December 2018. A full-scale experimental plan to assess oral susceptibility of sheep to CWD isolates from Nordfjella has been detailed in a proposal submitted to the Norwegian Research Council in September 2018. Evaluation of this proposal will be ready by the end of 2018. Thus, although preliminary data will be available from the ongoing experiment soon, it is likely that a more comprehensive analysis is still 2-3 years ahead.

1.3.3 Samples of rectal tissue and faeces from sheep that have grazed in Nordfjella Z1

Rectal tissues, including sub-mucosal lymphoid tissue and feces, from 800 sheep that had grazed in the Fødalen area in NF-Z1 were sampled in September 2018. These animals (mostly 7-9 months old lambs) were transported directly from Fødalen to slaughter. The Fødalen area is considered relevant, because of high densities of sheep and since this area was also frequently visited by reindeer (Figure 3-4 and Figure 3-10). Analysis of these samples, which will include highly sensitive methods like RT-QuIC, awaits evaluation of the aforementioned research proposal (see 1.3.2), since they are costly and time-consuming. Still, some results from such analysis should be available by mid-2019.

Data from these experiments will provide some information concerning the susceptibility of sheep to the Nordfjella CWD strain and the potential role of sheep as silent carriers of this prion strain.

1.3.4 *PRNP*-genetic analysis of cervids in Fennoscandia

For prion diseases such as classical scrapie and CWD, there can be significant differences in disease susceptibility among animals. The primary genetic element that governs this is the gene that encodes the prion protein (*PRNP*), in which several disease modulating polymorphisms have been described. Characterizing such genetic variation in European cervid populations is therefore important. A project is ongoing in which *PRNP* genetic variation will be characterized from all major Norwegian cervids. Data from the Nordfjella outbreak will soon be submitted for publication. Preliminary data show that there is a high degree of *PRNP* genetic variation within the Nordfjella reindeer population and that their CWD susceptibility varies accordingly.

1.3.5 Supplementary salt licks as a potential transmission hot spot for CWD prions

Samples of soil from selected supplementary salt licks in NF-Z1 and NF-Z2 were collected in 2017 and 2018 in a project funded by the Norwegian Environmental Agency and performed by NINA. The sampling is planned to continue in 2019 and, depending on results, in the years to come. The aim of the project is to elucidate to which degree the use of man-made salt licks leads to increased risk of transmission of prions and parasites between species. The study areas are the wild reindeer areas of Nordfjella, Knutshø and Forolhogna. Images and video taken by wildlife cameras mounted at the salt licks are used to show how often the licks are visited by different species and which behaviour the animals display there. The footages also show how efficient the exclosures are in preventing use of the licks by cervids. Position data from radio-collared reindeer will be used to show which salt licks are visited by reindeer and to model how the licks affect area use and movements of the animals. The soil samples from Nordfjella will be analysed for CWD prions by Prion Misfolding Cyclic Amplification (PMCA) and for parasites by traditional parasitological methods and Environmental-DNA (eDNA) techniques. In addition, the soil characteristics including content of different minerals will be determined. PMCA will not be performed on samples from the two other areas. Preliminary data show that some of the salt licks in NF-Z1 are frequently visited by red deer, and that these animals easily get in and out of the slots in the fences (Figure 1-6). Images from NF-Z2 show that reindeer, at least those with relatively small antlers, also get in and out without problems (Figure 1-5).

2 Literature search

A literature search was performed (date) in SCOPUS, PubMed and ISI Web of Science Core Collection using following search strings:

- SCOPUS search ("chronic wasting disease" AND "environmental persistence" OR "soil" OR "silent carrier" OR "salt lick"). Search returned 60 results.
- PubMed search
 - (chronic wasting disease[Title/Abstract]) AND environmental persistence[Title/Abstract]. The search returned 5 results.
 - ((chronic wasting disease[Title/Abstract]) AND scavenger[Title/Abstract]). The search returned 4 results.

There were no restrictions on date of publication.

2.1 Relevance screening

The titles of all hits were scanned and, if relevant, the abstracts were also inspected. Articles were excluded if they did not relate to the terms of reference. The reference lists in selected citations were scrutinized to identify additional articles or reports that had not been identified by the SCOPUS and PubMed searches.

3 Hazard identification and characterisation

3.1 Environmental reservoirs

Prions can persist for a long time in the environment. This, and the phenomenon of silent carriers (see below), is fundamentally important for understanding the epidemiology of infectious prion diseases such as typical CWD (Almberg, Cross, Johnson, Heisey, & Richards, 2011; DeVivo et al., 2017; Edmunds et al., 2016).

It has long been known that prions can bind to soil and remain infectious for years (Brown & Gajdusek, 1991; Seidel et al., 2007) and it has been hypothesized that soil can serve as a reservoir of prions (Johnson et al., 2006; Schramm et al., 2006; Seidel et al., 2007), though the nature of the association between prions and soil remains incompletely understood (Christen B. Smith, Clarissa J. Booth, & Joel A. Pedersen, 2011). Especially clay minerals are

suspected to bind prions efficiently and facilitate their uptake in the gastrointestinal tract, thereby increasing their infectivity (Wyckoff et al., 2016).

Prions also bind to dust and has been demonstrated to be present in airborne dust up to 50 meters from the barns where scrapie infected animals were kept (Gough, Baker, Simmons, Hawkins, & Maddison, 2015). An experiment where low-particle size dust, made from a mixture of clay and brain homogenate from CWD infected animals, was sprayed into the nostrils of white-tailed deer (*Odocoileus virginianus*), indicated that prions bound to dust and inhaled are infectious (Nichols et al., 2013).

Experiments indicate that grass can absorb infectious prions (hamster adapted strain) from the soil and transport these to the stem and leaves which become infectious in experimental settings (Pritzkow et al., 2015), suggesting that plants might serve as vehicles of disease transmission and that this should be taken into account in risk assessments. If infective doses of prions should be present in some alpine plants, the longevity of these can provide a potential for very long persistence.

The same researchers also showed that materials such as wood, rocks, plastic, glass, cement, stainless steel and aluminium (but not brass) efficiently bound, retained and released hamster-adapted 263K scrapie prions, and that hamsters housed together with contaminated materials of these kinds developed disease (Pritzkow et al. 2018).

Studies in deer (*Odocoileus* spp.) and elk (also called wapiti, *Cervus canadensis*) in North America show that infected animals excrete CWD prions to the environment with saliva, urine and faeces (Haley, Seelig, Zabel, Telling, & Hoover, 2009; Henderson et al., 2015; Pulford et al., 2012) months before the appearance of clinical symptoms. Especially saliva and faeces seem to contain infective prions relatively early post infection (Haley & Hoover, 2015), while prions appear in urine at a later stage. As discussed in the Phase II report (VKM, 2017b), quantifying infectivity levels in bodily excretions and determining their role in CWD epidemiology is not easy. Nevertheless, congregation of many animals in small areas for prolonged periods and/or by repeated visits, will allow gradual build-up of local environmental infectivity derived from all primary excretions. The potential role of such an area as a hotspot for disease transmission will therefore be a result of the total level of infectivity at the site, regardless of the primary sources of infectious prions.

Unfortunately, there are no established methods available for accurate quantification of environmental CWD infectivity-loads that could be used to classify potential hotspots for disease transmission. Therefore, in Nordfjella, detailed knowledge of the area-usage by wild reindeer, particularly the infected population in Zone 1, provides a guideline for evaluation and classification of putative hotspots for disease transmission.

In addition to secretory and excretory fluids, CWD prions are found in lymphoid tissues and blood at an early stage of the incubation period, and as the disease develops, the levels are increasing in both peripheral tissues and the brain (Hoover et al., 2017). Carcasses of animals that die of any cause during the development of CWD, can hence act as a point-

source of environmental contamination. Consequently, where there are cervids infected with CWD, we have to expect that the environmental contamination will be present at varying levels.

Low levels of CWD prions have been detected in environmental water samples from endemic areas during snow melt (Nichols et al., 2009). The prions were detected in both raw water and flocculate, but not in processed drinking water from the same source, indicating that the prions were bound to soil particles flushed off the land during this period of increased run-off. BSE and scrapie agents kept in phosphate-buffered saline and waste-water under laboratory conditions for eight years were shown to retain some degree of infectivity (Marin-Moreno et al., 2016). This suggests that prions can persist in natural water sources, but it is not known how long they will retain their infectivity under natural conditions.

Plummer and co-workers (2018) examined samples from the vicinity of eleven mineral licks utilized by white-tailed deer in an area of Wisconsin with a CWD-prevalence of 6 – 19% in adult deer. The samples consisted of eleven soil samples, nine samples of water from undisturbed rain puddles and eight samples of water from disturbed rain puddles, i.e. puddles where the researchers had stirred up sediment from the bottom. The samples were analysed with mb-PMCA (microplate-based prion misfolding cyclic amplification), which is regarded as a highly sensitive method for detection of prions. Seven of eleven soil samples, four of nine undisturbed water samples and two of eight disturbed water samples had close to detection limit of prions. CWD prions were also found in six out of ten faecal samples collected near the mineral lick with highest levels of CWD prions in soil and water samples. These findings confirm that the examined mineral licks were contaminated with CWD prions and that they might be important sources for environmental transmission of CWD. The authors suggested that both deposition of saliva during soil ingestion and faecal deposition contributed to the contamination in and around the mineral licks.

An area contaminated with prions is, however, not believed to stay contaminated forever. Prions do degrade and lose infectivity or otherwise disappear from the part of the environment where they are accessible for animals (S. E. Saunders, Bartelt-Hunt, & Bartz, 2008). Repeated drying and wetting (Yuan, Eckland, Telling, Bartz, & Bartelt-Hunt, 2015), heavy rain and run-off during snow melting (Nichols et al., 2009), repeated freeze-thaw cycles (C. B. Smith, C. J. Booth, & J. A. Pedersen, 2011), microbial and enzymatic degradation (Rapp, Potier, Jocteur-Monrozier, & Richaume, 2006; Rodriguez, Bennett, & Johnson, 2012; C. B. Smith et al., 2011), high concentrations of soil humic acid (Kuznetsova, Cullingham, McKenzie, & Aiken, 2018) and presence of natural inorganic oxidants (Russo et al., 2009) can reduce prion infectivity. This is described in chapter 3.3 in VKM Report 2017:9.

3.2 Transmission of prions and the transmission barrier

Prion diseases can be transmitted experimentally within or between species by exposing recipient animals to infectious donor material (typically brain material from an animal with

prion disease) through inoculation, most frequently, into the brain (i.c.), the peritoneal cavity (i.p.) or the gastrointestinal tract (oral inoculation, o.i.).

Normally, prion diseases transmit poorly from one species to another (sometimes zero transmission) and only after administration of very high doses of infectious material into the brain. The attack rate is often low and those animals that develop clinical signs do so after prolonged and variable incubation periods. Secondary (and subsequent) passage in the same recipient species, with material from cases derived from primary transmission, normally occur more efficiently, with shorter and less variable incubation periods. After further sub-passages, the attack-rate reaches 100% and no further reduction of the incubation period occurs – the prion strain is now considered to be *adapted* to its new host. The increased attack rate and shortened incubation period from primary (between species) to subsequent (within species) transmissions, was coined the “species barrier” (Pattison, 1965).

The nature of this barrier has been comprehensively investigated. Transgenic mice expressing Syrian hamster and mouse *PRNP* genes were inoculated with either hamster or mouse-adapted prion of scrapie origin. The animals developed clinical signs and pathological changes characteristic of prion disease, but analysis revealed that when mice were inoculated with hamster-adapted prions, only the hamster PrP was misfolded and vice versa in mice inoculated with mouse-adapted prion strains (Prusiner et al., 1990). These, and subsequent studies led to the notion that the species barrier predominantly consisted of differences in the primary structure of interacting PrP molecules.

Similar observations have been done in studies of classical scrapie in sheep, where a profound *PRNP*-genetic influence on disease development has been observed. Some alleles (Prion proteins), such as V₁₃₆R₁₅₄Q₁₇₁, displayed very high susceptibility, whereas animals homozygous for the A₁₃₆R₁₅₄R₁₇₁ allele (two amino acids different from the former) were almost completely resistant (Goldmann et al., 1990; Goldmann, Hunter, Smith, Foster, & Hope, 1994; Tranulis, 2002; Tranulis, Osland, Bratberg, & Ulvund, 1999). Further studies with transgenic mice, demonstrated that homotypic transmissions were highly efficient – i.e. when the recipient transgenic mice express PrP molecules identical to that of the infectious donor.

Accordingly, it was observed that mice expressing human PrP showed no species barrier towards transmission with prions derived from human cases of sporadic CJD. However, they showed a significant barrier towards transmission from human cases of variant CJD, although, in both instances, the donor and recipient PrP sequence were identical (Collinge et al., 1995; Collinge, Sidle, Meads, Ironside, & Hill, 1996). The latter observation suggested that structural features embedded in the prion strain itself could influence transmission properties beyond the primary structure of PrP. This notion was further substantiated during the BSE epidemic with what appeared to efficient transmission of disease to exotic ruminants and felids, such as cheetahs, despite significant differences in PrP primary structures (Collinge et al., 1996). Another fascinating example is the bank vole (*Myodes glareolus*), which appears to propagate prions from a variety of species, including humans very

efficiently (Watts et al., 2014). Importantly, bank voles were also highly susceptible to North-American strains of CWD, with 100% attack rates and relatively short incubation periods (Di Bari et al., 2013).

We can conclude that both the *PRNP*-genetics *and* the prion strain are important for transmission properties.

The route of infection can also play an important role. In most studies of experimental transmission, intracranial inoculation has been used because it is much more efficient, in some instances more than 100 000 times more efficient, as judged by infectivity titers (Kimberlin & Walker, 1978).

Oral inoculation experiments in rodents (Lasmézas et al., 1996; McBride & Beekes, 1999; McBride, Eikelenboom, Kraal, Fraser, & Bruce, 1992), sheep (Heggebo, Press, Gunnes, Gonzalez, & Jeffrey, 2002; Heggebo et al., 2000; van Keulen et al., 1995; van Keulen, Schreuder, Vromans, Langeveld, & Smits, 2000), and with CWD in cervids (Fox, Jewell, Williams, & Miller, 2006; Sigurdson et al., 2002; Sigurdson et al., 1999), have, however, demonstrated a complex interaction between the prion agents and the peripheral lympho-reticular system along the gastrointestinal tract.

Prions can cross the mucosal surfaces and, to a varying degree, propagate in lymphoid germinal centers. In classical sheep scrapie, CWD in cervids and vCJD in humans, this clinically silent lympho-reticular phase is prominent feature of the disease. In the case of BSE in cattle, however, this phase of the disease is hardly detectable, although after oral inoculation of BSE agent in cattle, prions were detected in the terminal portion of the ileum (Terry et al., 2003). Interestingly, oral inoculation of BSE in sheep revealed a prominent presence of the agent in peripheral lymphoid tissues (Foster, Parnham, Hunter, & Bruce, 2001). Thus, the “lymphotropism” of the BSE agent appeared to increase upon transmission to sheep. This was also the case after oral transmission of BSE to primates (Herzog et al., 2004) and, as mentioned, in human cases of vCJD (Hill et al., 1999; Wadsworth et al., 2001).

This implies that the degree of lymphoid propagation of a prion is not dictated solely by the prion strain, but through an interaction with the infected host and the route of infection.

Thus, an animal can be infected and propagate prions in peripheral lymphoid tissues (and possibly other organs) without any clinical signs for a long time. Indeed, it was noted some 40 years ago that, in extreme cases, the incubation period of a prion disease could exceed natural lifespan (Dickinson, Fraser, & Outram, 1975). In a series of studies, in which mice were inoculated with hamster-adapted scrapie strains (236K and Sc237), it was observed that although the mice remained completely healthy (attack rate 0 %), peripheral tissues and brain of the mice contained significant levels of infectious prions (Hill et al., 2000; Kimberlin & Walker, 1978; Race, Raines, Raymond, Caughey, & Chesebro, 2001). This was an important finding, since it demonstrated that in assessment of prion transmission, clinical disease was not always a reliable readout. Some animals could remain healthy for a normal

lifespan, while still actively replicating prions in peripheral tissues as subclinical carriers. For an early review of this phenomenon see (Hill & Collinge, 2003).

It was also noted that a subclinical carrier state could be induced experimentally in transgenic mice by the inoculation of very low doses of infectious material (Thackray, Klein, Aguzzi, & Bujdoso, 2002). Analysis has shown that the incubation period of experimental prion disease decreases linearly with logarithmic increase in infective doses, for doses at the ID50 level or higher, but less consistently for doses below ID50 (Fryer & McLean, 2011). The same models also showed that doses 1000 times below the ID50 could produce disease, indicating that a lower threshold for a “safe” dose of prion infectivity seems unlikely.

In summary; the term “species barrier” of transmission of prion disease should be replaced by the term “transmission barrier” since it is influenced by elements beyond *PRNP*-genetics (similarity in primary structure of PrP), namely the prion strain itself, the infective dose and route of infection. Furthermore, transmission efficiency cannot rely solely on observation of clinical disease since, in some instances, animals can propagate prions and generate and shed infectivity in the absence of clinical signs (see below).

As already mentioned, these phenomena are important when evaluating contagious prion diseases, such as CWD among cervids, but also for assessment of the potential zoonotic threat posed by CWD.

3.3 Carriers of disease

Any animal that in one way or another can contribute to the transmission of CWD in Norway will in the following text be denoted a “Carrier”, and grouped as follows:

1. *Clinical cases*
2. *Silent carriers (no clinical signs).*
 - a. *Preclinical*
 - b. *Nonclinical*
 - c. *Latent*
 - d. *Passive*

Importantly, an animal can carry and shed CWD prions at levels below detection limits or in a matrix that makes detection difficult, such as in faecal deposits. Thus, some carriers will most likely remain undetected. In the following description, the four groups of silent carriers will be in focus.

3.3.1 “Preclinical carriers”: Animals shedding infectious prions during the incubation period

A preclinical carrier is an animal in the incubation phase of CWD, i.e. it has been exposed to an infective dose of prions and become infected, but clinical signs are not yet visible. Per

definition, preclinical carriers will develop clinical signs of disease following the incubation period and eventually die of CWD, unless they die of other causes.

As illustrated in the figure below, preclinical carriers can propagate prions in peripheral tissues, which leads to a gradual build-up of infectivity and increased probability of prion release. Data have shown that such animals can shed prions in faeces, urine and saliva for a prolonged period (months) before clinical disease becomes apparent, and that this is important in CWD epidemiology (Di Guardo & Marruchella, 2010; Haley, Mathiason, Zabel, Telling, & Hoover, 2009; Haley, Seelig, et al., 2009; Kurt & Sigurdson, 2016; Kuznetsova, McKenzie, Baner, Siddique, & Aiken, 2014; Miller & Williams, 2003; O'Rourke et al., 2003; Sigurdson, 2008).

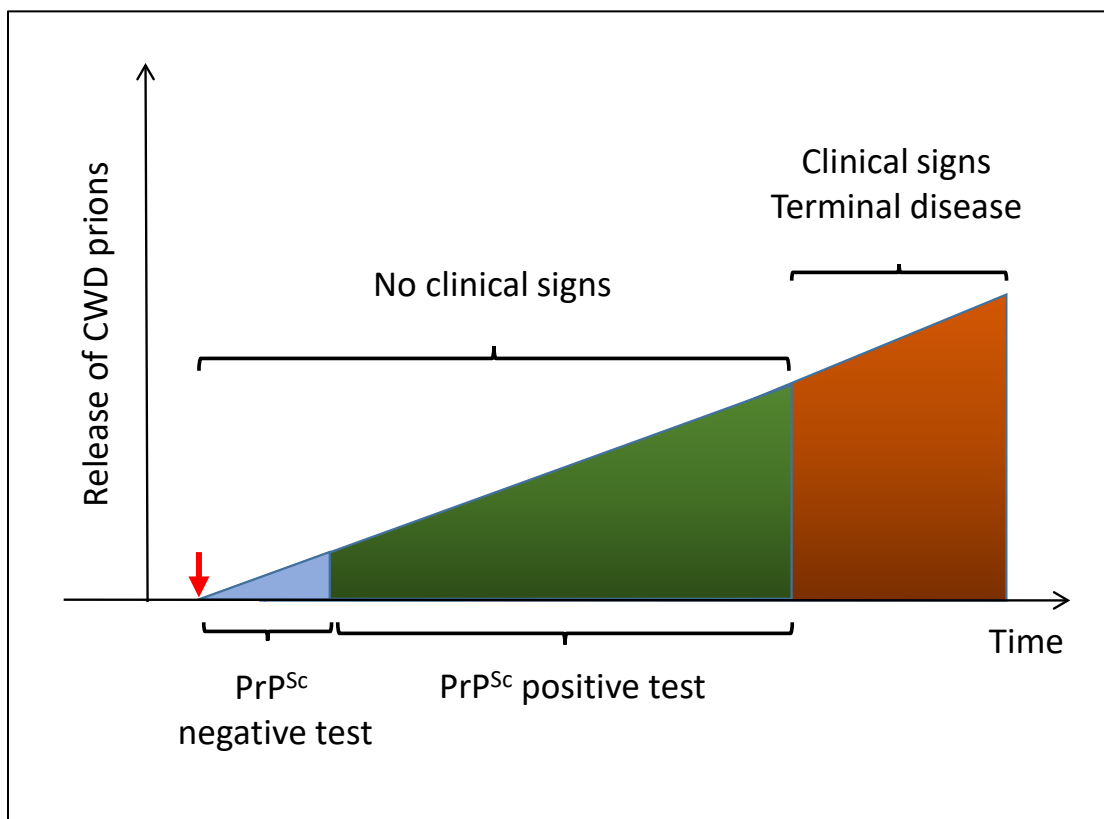


Figure 3-1. Schematic illustration of a preclinical carrier situation. The red vertical arrow indicates the time of infection. After a certain time, PrP^{Sc} levels in peripheral tissues have reached a level sufficient for detection and prions can be shed to the environment. Whether such shedding is continuous or intermittent/variable is not taken into consideration. As the prion agent reaches the brain, clinical symptoms can gradually develop. The slope and shape of the line and duration of the preclinical and clinical phases probably vary significantly according to factors such as the host susceptibility, infective dose and route of infection in addition to the prion strain itself. This illustration is not intended to describe either the rate or dynamics of the three phases.

As far as we currently know and based on knowledge from North America supplemented with pathological findings in the reindeer cases from Nordfjella, the course of clinical CWD in Norway should be anticipated to include a lengthy incubation phase under which the animal

might shed infective prions before clinical disease ensues. Consequently, most of the CWD-positive animals in Nordfjella presumably were pre-clinical carriers. We do currently not know to which degree red deer, moose and roe deer are susceptible to the strain from Nordfjella, but based on experiences from North America, it can be presumed that these species also are susceptible to infection and will go through an incubation phase as preclinical carriers.

It should, however, be noted that deer with moderate susceptibility, for instance modulated by *PRNP* genetics, could upon exposure to very low infectious doses experience a lengthy incubation period and die of other causes before developing CWD – and thus, for all practical purposes, *appear* as nonclinical carriers (see below).

3.3.2 “Nonclinical carrier”

The nonclinical carrier, illustrated in the figure below, denotes a hypothetical carrier state in which the CWD agent propagate, reach detectable levels in lymphoid tissues and is shed with excreta (Mathiason, 2015). The propagation occurs slowly and clinical CWD does not occur during the normal lifespan of the animal. We are not aware of any scientific studies showing that nonclinical carriers, as defined here, play a role in CWD epidemiology. However, this phenomenon has not been studied much and, as previously discussed, similar carrier states have been observed in experiments. The nonclinical carrier state should be regarded as hypothetical, while recognized as a knowledge gap.

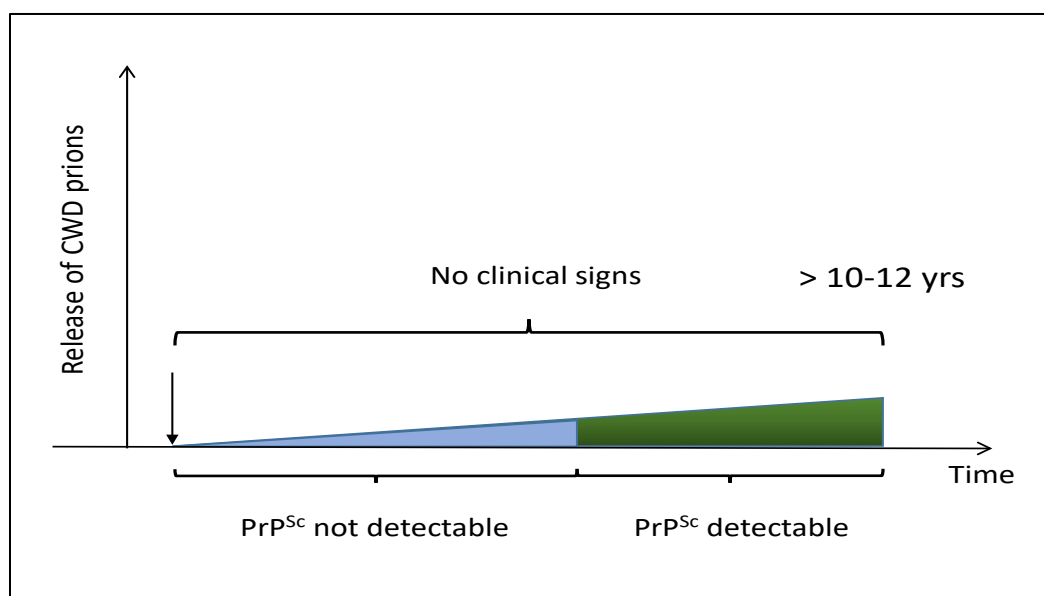


Figure 3-2. Schematic illustration of a nonclinical carrier of CWD infectivity. This hypothetical animal is infected by CWD and propagates prions slowly, but sufficient to reach detectable levels in peripheral tissues and for release to the environment. The slope of the line and duration of the phases can vary. Clinical disease does, however, not occur during the animal's normal lifespan. It is assumed that the level of released prions from such an animal is much lower than from a preclinical carrier. Whether sheep exposed to CWD prions can function as nonclinical carriers is currently under investigation. This illustration is not intended to describe either the rate or dynamics of the two phases.

Finally, it cannot be excluded that livestock, which are presumed to have very low susceptibility towards CWD, can harbor and shed CWD prions from peripheral tissues, like those of the gastro-intestinal tract, and thus function as nonclinical carriers. With regard to sheep, this question is currently under investigation.

In an experiment with intracranial inoculation of CWD in sheep, one clinically healthy animal was euthanized 72 months (six years) after inoculation, and turned out to be positive for PrP^{Sc} (Hamir et al., 2006). This demonstrates that even after intracranial inoculation of a high dose the transmission barrier apparently turned this animal into a carrier that could have developed clinical signs (preclinical) or remained healthy (nonclinical). Interestingly, in the same study, another sheep with a different *PRNP*-genotype (VRQ/ARQ) developed clinical disease three years after inoculation, suggesting that *PRNP*-genetic modulation could be important. In the ongoing oral inoculation experiment at NMBU, mentioned above, sheep with the VRQ/VRQ *PRNP*-genotype have been selected due to their high sensitivity towards classical scrapie prions.

It should be emphasized that even though animals with low susceptibility in some experiments have been shown to replicate prions without developing symptoms, the amount of prions shed must be regarded to be a lot higher from susceptible animals than from less susceptible.

3.3.3 “Latent carriers”:

Latent carriers comprise animals that are infected and in which prions are propagated but not shed to the environment in epidemiologically relevant doses. Transmission to other individuals does not occur unless infected tissue from the latent carrier is eaten or otherwise transmitted directly to a new individual or indirectly through the environment, for instance by decaying carcasses. Cattle with BSE might serve as an example of a latent carrier, since BSE is not transmitted directly between animals in a herd but can be transmitted to bovids and other species (see above) with meat products from infected animals. There is to our knowledge, however, no reports showing that “latent carriers” contribute to dissemination of CWD prions, but the possibility cannot be excluded.

3.3.4 “Passive carrier”:

The passive carrier is an animal that has been exposed to CWD prions but without any uptake of the agent over mucosal surfaces. There is no prion propagation and amplification in these animals and prions cannot be detected in any tissues, but low levels might be present in the content of the gastro-intestinal tract, including faeces, or on body surfaces (skin, hooves, wool, antlers etc.), from which the prions can be liberated. This group also includes carnivores, scavengers, rodents, insectivores, insects and birds that can distribute potentially infectious material from a carcass. The relative importance of passive carriers in CWD epidemiology is unknown, but it may pose a risk to geographic spread. Technically, any

animal exposed to contaminated material NF-Z1, including man, can act as a passive carrier (Figure 3-3).



Figure 3-3. Golden eagle (*Aquila chrysaetos*) feeding on the remains of a reindeer carcass in Nordfjella Z1. Some ravens (*Corvus corax*) are also visiting the carcass (left). These are examples of animals that can potentially distribute CWD infectious material as passive carriers. Photo: Olav Strand NINA.

Table 3-1. Summary of carrier types, susceptibility and prion releasing

Category	Susceptibility	Agent taken up and propagated		Agent release	Host species	Progression to clinical disease
		Lymphoid	Brain			
Preclinical carrier	High/moderate	High level	No	High level, prolonged	Cervids	Yes
Nonclinical carrier	Low/moderate	Detectable	Not during normal life span	Unknown, probably very low	Any animal with incubation period exceeding normal life span	No
Passive carrier	Extremely low	No	No	Very low and transient	Livestock, carnivores, scavengers, other	No
Latent carrier	Unknown	Very low	No	Not from live animal	Unknown	No

3.3.5 Non-biological mechanical transport of CWD prions

This refers to either surfaces directly contaminated with excretions containing CWD-prions or mud, soil and faeces from animals contaminated with CWD-prions and CWD-prions bound to surfaces of equipment, mobile installations or vehicles like metal fences, mineral lick stands/holders, boots, trucks, cars, tractors, snowmobiles etc. Vehicles used for transportation of animals from NF-Z1 are particularly relevant.

3.4 Cervids in Nordfjella

Background information on the cervid population in the Nordfjella region, its distribution and population development, movements and migrations are described in detail in Appendix I.

The distribution of cervids around Nordfjella fluctuate over time due to long-term recolonization processes from a previously low population density, immigration from neighboring populations experiencing high densities and anthropogenic factors including infrastructure. Seasonal migrations and shifts in habitat selection are also important. The patterns of cervid distribution are therefore in part driven by factors outside of the Nordfjella area.

3.4.1 Wild reindeer

Reindeer are to a large extent living above the tree-line and populations are therefore distributed within a matrix of mountainous and forested habitats where human infrastructure and disturbance also act to hinder reindeer movements between populations. Management is to a large extent adapted to these distribution patterns and administrative management borders do to a large extent follow the biological borders of each population and not municipality borders. At present, wild reindeer are living in more or less closed populations where management by harvest aims to keep populations at predetermined sizes.

3.4.1.1 Large scale drivers and factors external to Nordfjella

Distribution of reindeer in the Nordfjella region has been influenced by several external factors during the last decades. Most importantly, when the population density peaked in the Hardangervidda mountain plateau, reindeer from this population migrated into Nordfjella. This occurred twice, first in the late 1960s, thereafter in the early 1980s. During the last migration, anecdotal information indicate that more than 10.000 animals moved northwards to utilize winter pastures in Nordfjella. This influx of animals led to dramatic changes in management policy and hunting quotas were increased considerably in order to reduce the population density to protect winter foraging areas from rapid depletion. These invasions also introduced wild reindeer to the mountains west of Nordfjella, and a new wild reindeer area was established in Raudafjell Mountains. However, as these populations later declined and finally disappeared, the Raudafjell wild reindeer area was discontinued in the early 1980s.

The highway RV7, crossing Hardangervidda, is a strong barrier to reindeer movement and hinders animals from the southern/central parts of the mountain plateau to move into the northern parts of the range (denoted as the HJ (Hardangerjøkulen) area in figure 3-4) and further into Nordfjella. RV7 was opened for winter passage by cars for the first time in 1982. The combination of this barrier and the reduced population size in Hardangervidda, declining from approximately 25.000 animals in 1984 to between 5.000 and 10.000 animals in total today, led to a situation where HJ and the southern part of Nordfjella (NF-Z2) held only a small number of reindeer during the 1990s.

Infrastructure has also influenced reindeer distribution and habitat use in Nordfjella (Nellemann, Vistnes, Jordhøy, & Strand, 2001; Strand et al., 2011). Firstly, the highway RV52 (see above) functioned as a barrier for movement between the wild reindeer in NF-Z1 and the semi-domesticated herds of Filefjell Reinlag (Filefjell Reindeer Breeders' Co-operative). Secondly, several hydroelectric water reservoirs and the road FV50 divided Nordfjella into two more or less separated populations. Thirdly, the railroad "Bergensbanen" that crosses the mountains between Hardangervidda and Nordfjella (Figure 1-1) has been regarded as a barrier to reindeer movements between Hardangervidda and Nordfjella.

In 2001 some reindeer migrated from Zone 1 to Zone 2 in Nordfjella, probably because of suboptimal foraging conditions in Zone 1 and high levels of human activity in the traditional pastures in the eastern part of Zone 1. Given that CWD might have been present in NF-Z1 for more than 15 years, the abovementioned migration underscores the importance of comprehensive surveillance of the reindeer population in NF-Z2.

Animal tracking with GPS-collars in Nordfjella started in 2007 and provided valuable data on distribution and habitat use in this population. Since 2007 a total of 53 animals have been collared (27 females in NF-Z2 and 26 females and 5 males in NF-Z1). Tracking data from these animals showed that reindeer in NF-Z2 utilized HJ on a regular basis (Figure 3-4). GPS data from collared animals in the Hardangervidda herd confirmed that reindeer in Hardangervidda are limited to habitats on the southern side of RV7, although animals from Hardangervidda occasionally and temporarily have crossed the road. For example, approximately 100 animals, including one collared female, made a brief winter excursion north of the highway in 2007. This, and similar groups of reindeer, crossed the road in periods of harsh weather during which the road was closed for ordinary car traffic. These flocks were later observed to return southwards to the central parts of Hardangervidda. Although none of the other radio-collared animals ($n = 148$ in Hardangervidda) and 27 in Zone 2) are known to have crossed the barrier at RV7, this highway cannot be regarded as an impermeable barrier for the reindeer. Most importantly, the collaring program has focused on female reindeer and we have far less knowledge about the males, which are known to have a more gregarious behavior and are more likely to disperse.

3.4.1.2 Local drivers in Nordfjella

Reindeer distribution in Nordfjella is also a result of drivers operating at a local scale. Among them, local population management is important, acting to modulate the already described fluctuations in population density. Management has also had substantial effects on population sex ratio, and probably also on habitat use.

The male segment of the reindeer populations was severely overharvested in the early 1980s and as little as 4-8 % of the populations was classified as adult males by the end of that period. Active management and sex and age specific harvest quotas has helped to restock the male segment to today's levels of 18- 23% (Solberg et al., 2017).

The sexual dimorphism, behavioral differences and reproductive strategies adopted by reindeer (see appendix) results in highly different energy budgets and consequently herd behavior and habitat use. Males suffer the highest energy expenditure in fall in connection with the rut, females in late winter and the first weeks after giving birth. Adult males are less prone to predation and have therefore a more gregarious habitat use. They form small male groups prioritizing nutritious habitats and gain of body mass through a large part of the spring and summer. Females with calves are far more prone to predation and form larger nursing groups searching for undisturbed habitats, balancing the risk between predation and access to early green pastures. If possible, females aggregate in remote calving grounds within short distance to habitats at low elevations with early green up.

GPS-data from the collaring program in Nordfjella reveals relatively large geographical differences in habitat use (Figure 3-4). Areas to the east are less used by reindeer, whereas central and western parts, in general, have a higher probability of being used. To some extent, we also see seasonal differences in habitat use. Of particular interest to the scope of this report is a general movement to the west and to low-lying habitats, in spring and early summer, bringing reindeer closer to habitats heavily used by red deer. In Zone 2, reindeer also have a pronounced shift to the north and west during summer, but, most importantly, uses habitats close to RV7 throughout the year. At the same time, reindeer in Hardangervidda have a seasonal shift in habitat use with an almost exclusive use of a limited habitat in the southern parts of Hardangervidda during summer (Strand, Jordhøy, Panzacchi, & Van Moorter, 2015). It therefore appears that a direct contact, and with that a potential direct transmission of CWD, between reindeer from Hardangervidda and Zone 2 is most likely to occur in late fall and winter. The presence of mineral licks in the areas where there is overlap, will however facilitate indirect transmission, and thereby increase the potential for transmission between the populations manifold.

Most notably, the collaring programs has largely included females, since there are practical difficulties associated with collaring of males, and that collaring can affect their welfare. Far less is consequently known about the dispersion of smaller groups of males between the different areas. Previously, when the Hardangerjøkulen (HJ) area held a substantial reindeer population, it was a good summer habitat for males. Males who had their summer grazing area here migrated each fall to Hardangervidda for the rut. According to anecdotal evidence,

this annual movement of males was halted during the late 1980's and early 1990's when the population size in the HJ area declined. It therefore seems prudent to document male behavior and migration between the HJ area and Hardangervidda south of RV7, since reindeer from Zone 2 are using the HJ area.

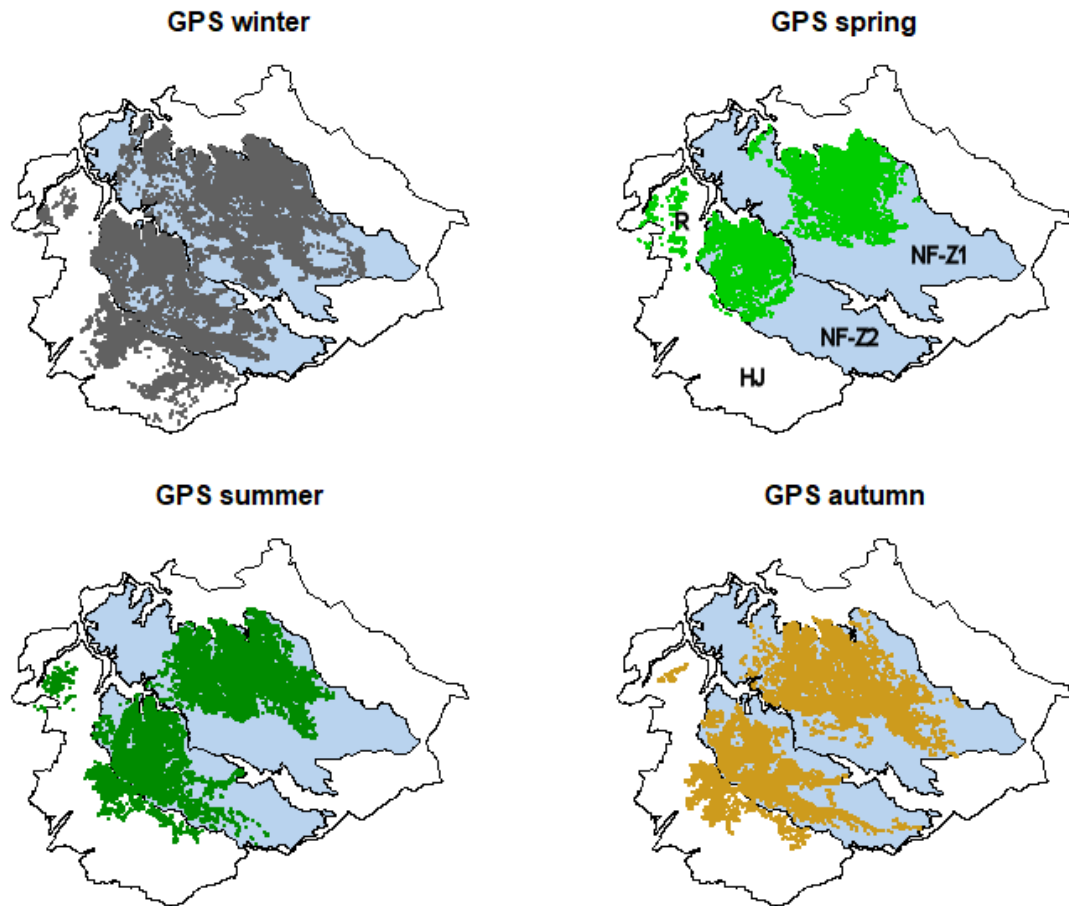


Figure 3-4. GPS data and reindeer habitat use in NF-Z1 and NF-Z2 in winter, spring, summer and autumn. Notably, the NF-Z2 herd utilizes areas south of the NF-Z2 area, in the northern parts of Hardangervidda around the glacier Hardangerjøkulen (HJ). During winter time, the animals go all the way down to RV7 (upper left). In addition, reindeer from NF-Z2 also wander westwards into Raudafjell and spend time there, especially in the spring and summer (R).

3.4.1.3 Translocation of reindeer from Nordfjella to other areas.

Similar to most wild reindeer areas in Norway, also Nordfjella has a history of domestic reindeer breeding. As wild populations increased in numbers during and after World War 2, herding became increasingly difficult, and reindeer herding enterprises were discontinued one by one. In Nordfjella, Hol austre tamreinlag ceased their semi-domestic reindeer operation in Hallingskarvet Mountains in 1982. Most animals were slaughtered, but some live animals were transported to other areas, among them Nya Idre sameby in Sweden. We do

not know how many animals were transported out of Zone 1 or to which destinations they were transported.

Until the autumn of 2016, a few semi-domestic reindeer were held at Geitryggghytta Tourist Cabin owned by The Norwegian Tourist Association, located on the border between NF-Z2 and NF-Z1 (Figure 3-5). Anecdotal information suggests that these were the property of the former manager of the lodge. The number of reindeer has varied between a few animals up to approximately 20. At large, these animals stayed close to the cabin, but in periods with higher reindeer numbers they also roamed over a larger area and most probably also into NF-Z1 (as the FV50 goes into a 3,3 km long tunnel at this point and there consequently is no obstacle for reindeer movement between the zones). Normally, keeping of semi-domesticated reindeer is prohibited in wild reindeer areas, and we are not aware that any dispensation was granted by the authorities. The last animals from this herd were, as far as we know, slaughtered in 2016, but anecdotal information indicates that animals have been transported to/from Geitryggghytta and other reindeer herds on several occasions.



Figure 3-5. Semi-domestic reindeer were kept as pets at Geitryggghytta Tourist Cabin for several years. The reindeer disappeared sometime after 2011, but their destiny is unknown to us. The pictures were downloaded from <https://geitryggghytta.dnt.no/bilder> the 27th of March 2017.

In 1994 and 1995, a total of 20 wild reindeer were captured in Zone 1 and translocated to the Lærdal – Årdal area. This was done to reintroduce a population there, since the reindeer population in that area had declined and disappeared around 1980-1985. Prior to this, Lærdal - Årdal had a viable wild reindeer population of about 350 animals. The disappearance was, again according to anecdotal information, a result of assimilation of the wild reindeer into a new semi-domestic herd established in the area in 1972. The restocking initiated in 1994-95 was successful and the population has counted approximately 270 animals during the last decades with a yearly harvest of 50 animals. Recent population surveys do however indicate a significant population decline, and harvest has been banned the last three years.

3.4.2 Semi-domesticated reindeer in Filefjell Reinlag

Filefjell reinlag was established in 1945 after the termination of Jøkulen rensselskab, which had a reindeer herd in Filefjell until that time. The company, owned by five inhabitants of the

area, use areas in nine municipalities and three counties for pasture. The winter herd size has been between 2700 and 3100 since the establishment. Since 1945, the herd is managed according to the Rørros-model, i.e. all male yearlings and most calves are slaughtered each autumn and the winter herd consist of approximately 2400 females (80%), 300 female and 300 male calves (20%) and 10 males. In the winter, the herd utilizes pastures far south in the area, in the mountains north and south of Vassfaret in the municipalities Sør-Aurdal and Flå (Figure 3-6). Before calving, which occur from the last part of April until around the 20th of May, the females are driven 125 km in areal line northwards to the north-eastern part of Filefjell, in Vang municipality. In some years, however, weather and snow condition cause the herd to move further west, to Oddedalen in Lærdal municipality. From around the 20th of May and through the summer and autumn, the herd grazes in the areas between the roads RV52 and E16 (Figure 3-7 and Figure 3-8).

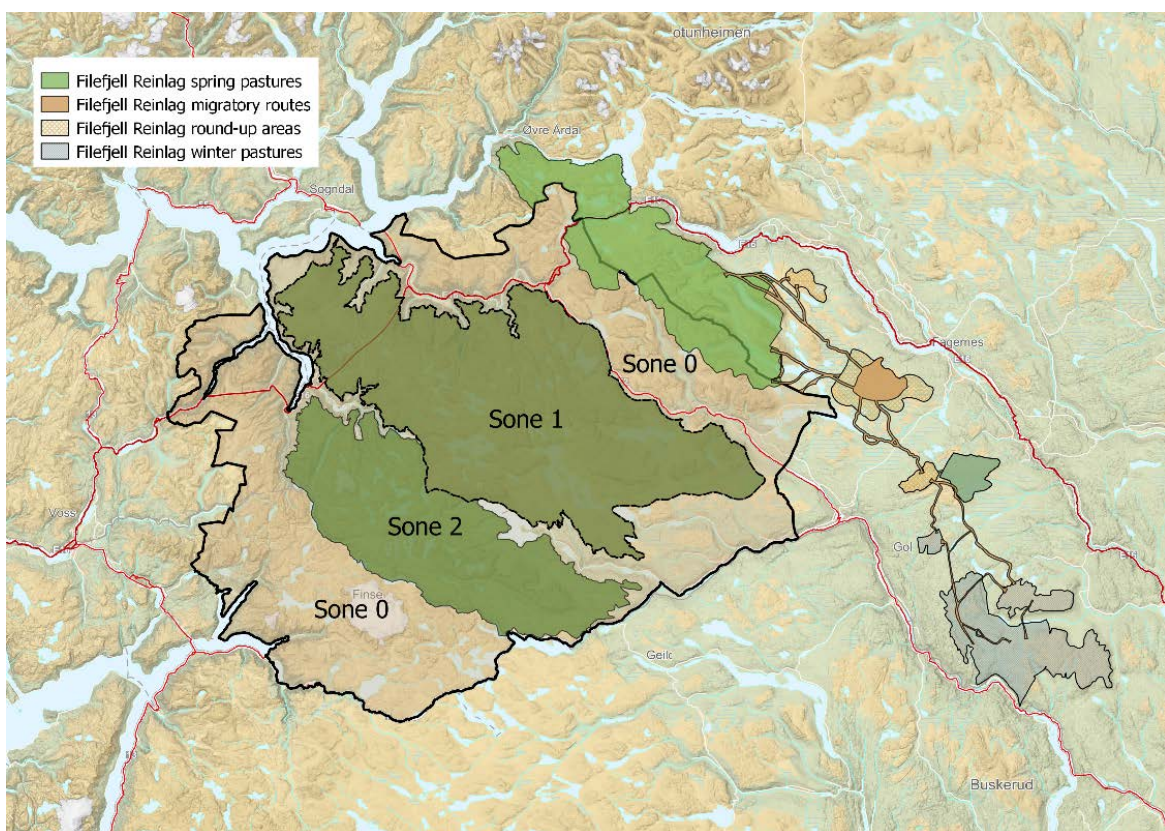


Figure 3-6. Pastures, migratory routes and round-up areas used by Filefjell reinlag in winter and spring. During winter, a herd of app. 3000 animals use areas relatively far south in Buskerud and Oppland counties (grey). During April, the animals are herded northwards along established migration routes. Calving occurs in April – May in the north-eastern part of the area (green). The green area north of E16 (red line), which is a part of Lærdals-Årdalsfjella, is currently not used by Filefjell reinlag. (Source: www.kilden.no)

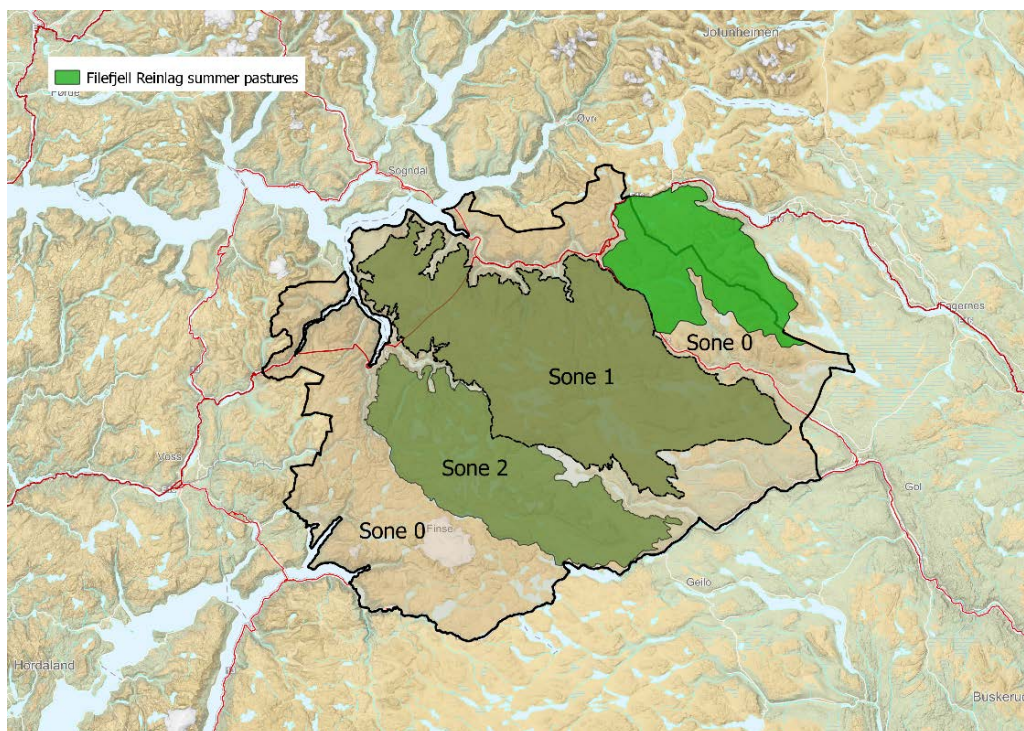


Figure 3-7. Pasture areas used by Filefjell reinlag ANS in summer. A herd of app. 5000 animals graze in the area between the roads E16 and RV52. Consequently, only the latter road has separated the Filefjell herd from the now eradicated herd in NF-Z1 and the areas that potentially were, and probably to some degree still are, contaminated with CWD prions. (Source: www.kilden.no)

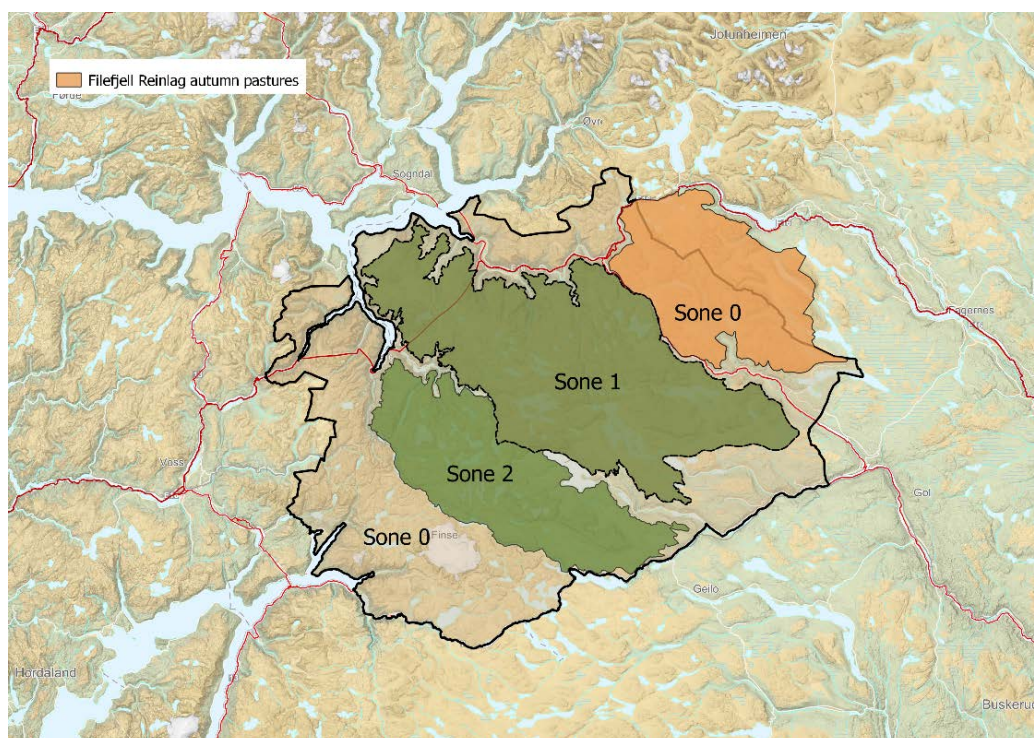


Figure 3-8. Pasture areas used by Filefjell reinlag ANS in the autumn, before the snow cover allows herding south-eastwards to the slaughter place. Also in this period, the Filefjell herd used and still

uses the areas along RV52, close to the north-eastern border of NF-Z1. In late autumn, salt sprinkled on the road to prevent icing attracts the animals to the roadsides. (Source: www.kilden.no)

The semi-domesticated reindeer consequently use pastures along the northern border of NF-Z1 during the green seasons. In this period, the herd is seen after daily by the herdsmen. When the road authorities spread salt on the roads in autumn to prevent icing, the reindeer often lick salt along the roadsides (Figure 3-9). As the snow arrives, the herd is driven eastwards and southwards into Nord-Aurdal municipality, where between 2000 and 2200 animals are slaughtered, and the remaining winter herd is herded back to the winter pastures.



Figure 3-9. Semi-domesticated reindeer owned by Filefjell reinlag lick salt on RV52 on the 7th of October 2018. The road authorities use salt to prevent icing forming on the asphalt. This road constitutes the border between NF-Z1 and the semi-domesticated reindeer area. (Image taken by Emilia Næsvik.)

Filefjell reinlag has responded to the presence of CWD in NF-Z1 by termination of their use of salt licks (they used to have 2000 kg mineral stones placed out on the pastures) and reducing their winter herd with 10% from 3318 in 2016/17 to 3050 in 2017/18 (pers. comm. Filefjell reinlag ANS). In 2017, the company has slaughtered all animals older than 9.5 years and tested them for CWD. Since the finding of the first CWD-positive reindeer in Zone 1, samples from well above 900 reindeer from Filefjell have been tested and diagnosed CWD-negative.

Before the eradication of the herd in NF-Z1, wild reindeer males from this herd relatively often crossed over RV52 to graze on the fresh grass of the southern slopes of Filefjell in the spring. Reindeer from Filefjell reinlag have sometimes crossed over to NF-Z1. The number of animals that crossed before 2016 is uncertain, but according to data provided by H. Skjerdal

in Aurland fjellstyre (Aurdal Mountain Management Board), between 0 and 8 animals has been shot during the wild reindeer hunt in NF-Z1 between 2008 and 2016. During the hunt in 2017, a herd of 23 and seven additional solitary semi-domesticated reindeer were shot in NF-Z1, while the reindeer herders have shot 32 animals in or on the border of NF-Z1 in 2018. The areas where the reindeer use to cross are now closed with 24 km of reindeer fence (see above). To ease herding and prevent that the reindeer wanders into NF-Z1, the company also has invested in 170 new radio collars (in addition to the 30 radio collars they had before).

3.4.3 Red deer

Red deer have higher population densities on the western side of the mountains (Figure 8-1 and Figure 8-2). This is reflected by high harvest densities of red deer in the municipalities Lærdal, Aurland and Årdal in Sogn og Fjordane County and Ulvik in Hordaland County. However, red deer have expanded markedly towards the east during the last decades and harvest numbers have increased considerably in the municipalities of Hol, Hemsedal, Ål and Gol surrounding the eastern side of Nordfjella. Still, the numbers and densities are low compared with those on the western side of the mountains.

Spring migration of red deer peaks the second week of May, while fall migration starts early September and lasts into November (Meisingset et al., 2018). Spring/summer migration is driven by early plant growth (Bischof et al., 2012), while onset of fall migration is partly triggered by onset of the hunting season as well as cold spells and snowfall later in the season (Rivrud et al., 2016).

The spatial overlap with the reindeer range of Nf-Z1 occurs mainly during early summer to late autumn and reaches considerable levels in Lærdal (Figure 8-3). Both migratory and resident red deer in Lærdal use the forested areas during early summer to late fall. Population densities are likely to be $<5/\text{km}^2$ based on harvest numbers in the west but may locally be much higher as red deer are social.

3.4.4 Moose

Moose have higher population densities on the eastern side of Nordfjella (Figure 8-1 and Figure 8-2). The municipalities of Hemsedal, Hol, Ål and Gol in Buskerud County have sizeable populations of moose (Figure 8-1 and Figure 8-2). Only a few moose are shot annually in the municipalities on the western side of Nordfjella, and the densities are unlikely to grow much.

Moose migrations are also driven by snowfall in the autumn and expansions in the spring, as the snow melts, although moose are less sensitive to snow compared with red deer and roe deer. Moose migrations are to a lesser degree driven by maturation of grass and herbs during spring than red deer migrations, due to their reliance on browsing trees and bushes.

The spatial overlap of reindeer in Nf-Z1 and moose was low based on existing data (Figure 8-3). However, moose that were selected for collaring were not recruited from areas close to Nordfjella. More anecdotal observations suggest that individual moose can spend quite some time browsing in particular willow in the low alpine areas of Nordfjella during summer (Mysterud, personal communication). But still, overlap between moose and reindeer is much less than for red deer-reindeer. The overlap is likely partly reduced due to reindeer being more sensitive to human disturbance, which is more frequent on the eastern side of Nordfjella. Population densities of moose are likely to be up to 1-1.5 per km² based on harvest numbers.

3.4.5 Roe deer

Due to severe winters and predation by lynx, the population size of roe deer around Nordfjella is low. There were shot 25 roe deer in Hemsedal in 2017, and for Hol, the maximum over the period was 55 in 2017. At such low numbers, the data from Statistics Norway are not reliable as they do not report numbers unless more than 25 roe deer are shot in a given municipality a given year. During 1986-2017, there were no roe deer registered shot in the municipalities west of Nordfjella (Lærdal, Aurland and Årdal in Sogn og Fjordane and Ulvik in Hordaland).

There is much less available information based on GPS-marked roe deer, though many roe deer were marked in the area when VHF collars were used (Andersen, Linnell & Aanes 1995; Mysterud 1999).

The spatial overlap of reindeer in Nf-Z1 and roe deer is estimated to be minimal. The empirical observation was based on few individuals (Figure 8-3). However, based on general knowledge about roe deer biology, the overlap with reindeer is unlikely to be important, considering the low population density of roe deer and their preference for valleys close to human agricultural areas.

3.5 Domestic animals grazing in Nordfjella

An overview of Norwegian sheep farming and its organisation is given in the Appendix II.

Fertile grasslands in the Nordfjella mountain range have, probably for centuries, been utilized as summer pastures for sheep and other livestock. Most farmers are local to the valleys and along the fjords surrounding the mountains. The municipalities of the NF-MA, i.e. Hemsedal, Ål, Hol, Eidfjord, Ulvik, Aurland and Lærdal had a winter population of 22.060 sheep in May 2017 (Statistics Norway). As each ewe in average gets close to two lambs (in 2017 the average number of lambs per ewe in this area was 1.82) this approximates 60.000 sheep released on pasture from farms in the seven municipalities. The overall number of sheep in this area has been relatively stable (Figure 3-9), with a 13% decrease since 1989. There are, however, large variations between the municipalities, spanning from 69% decrease in Eidfjord and 44% in Aurland to a 21% increase of the sheep numbers in Hemsedal. In the

same period, the number of farms with sheep has decreased with 54%, which implies that the average winter herd size in the area increased from 40 to 75 animals (Statistics Norway). Not all these sheep graze in the Nordfjella wild reindeer area, however, as also the mountain areas within the rest of the NF-MA are used for grazing. In Lærdalen sankelag (Lærdalen Sheep Herding Co-operative), for example, only 41.5% of the sheep were released in the Nordfjella wild reindeer area in 2017, while 12 and 46% were released in the mountains (Figure 3-10) east and north of the Lærdalen valley, respectively (Lærdalen sankelag. Annual report (Årsmelding) 2017).

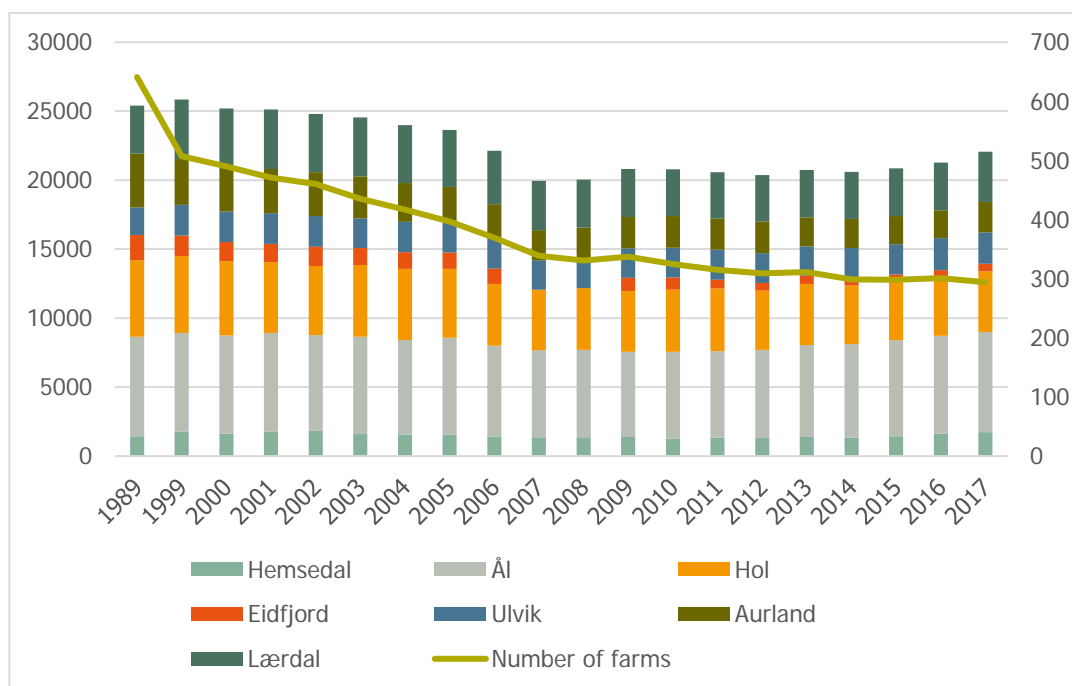


Figure 3-9. The number of sheep over 1 year of age housed in farms of the municipalities in the Nordfjella area (left vertical axis), has been relatively stable, showing a decrease from 25.403 in 1989 to 22.060 in 2017 (-13%). This equals around 60.000 ewes and lambs on pasture during summer (since the number of lambs per ewe averages 1.8). In the same period, the number of farms with sheep (right vertical axis) decreased with 54%, which implies that the average winter herd size in the area increased from 40 to 75 animals (Data from Statistics Norway).

In NF-MA pastures are divided between 16 such “beite-/sankelag” (sheep management co-operatives) organizing the clear majority of the sheep (figure: map from www.kilden.no) but with large variations in sheep number, area used, sheep density and management policy. Some of these beite-/sankelag consist of only local farmers, while others are so-called “drifter” which accept and take the responsibility for supervision of sheep herds from farms up to 200 km away during the pasture season. The density of sheep in some of the most fertile areas has been high. In 2013 the beite-/sankelag called Fødalsdrifta reached a record of 100 animals per square km accessible area (accessible area is the total area minus sea, lakes, agricultural area, built area, glaciers and bare rock). In the western beite-/sankelag, however, the density is as low as 6 animals per square km, giving an average density of around 15 sheep per sq. km accessible area. In 2017 the beite-/sankelags released a total of

57.709 sheep into an area of 3.757 sq. km accessible areas at least partially overlapping Nordfjella wild reindeer area (Figure 3-10 and Figure 3-11). In addition, 3.999 sheep were released south of Hardangerjøkulen Glacier, but north of the road RV7 in Eidfjord municipality, 3.207 in NF-Z0 areas of Ulvik and 697 in Flåm Vestfjell mountains west of Flåmsbana railroad in Aurland, making the total number of sheep released in the NF-MA in 2017 65.612.

The proportion of sheep that originate from farms outside the Management Zone (and consequently to some degree might be transported back and put on home pasture) is currently not established due to time and resource constraints but can be calculated by compiling data from several governmental registries (pers. comm. Michael Angeloff, NIBIO). One of these registries (Produksjonstilskuddregisteret) also contain information about the address and owner of the individual farm, so that the exact origin of the sheep can be provided. This is, however, not publicly available information due to personal protection regulations. However, the difference between number of sheep and lamb that participate in organised beite-/sankelags and the number of sheep and lamb released from farms within in the municipalities (Hemsedal, Ål, Hol, Eidfjord, Ulvik, Aurland and Lærdal), provide a rough indication of the number of sheep and lamb that originate from other areas (Table 3-2). A positive number indicate that at least that many sheep originate from municipalities other than the municipality in question, while a negative number either can indicate that there is a proportion of sheep farms that do not participate in organised beite-/sankelag, or that their sheep and lamb are sent to pasture in other municipalities. Based on this simple estimate, at least 11.644 sheep/lamb originate from municipalities outside the management area.

Table 3-2. Difference between total number of sheep and lambs released in the beite-/sankelags of the municipalities in Nordfjella and the number of sheep and lambs released to pasture from farms in the same municipalities. NF-MA only comprise parts of Ål, Hol, Eidfjord and Ulvik, explaining the difference between number of sheep released in the NF-MA compared with the sum of sheep and lamb released in the municipalities. (Data from the registries Organisert beitebruk (OBB) and Produksjonstilskuddsregisteret (PT) collected by Michael Angeloff, Norwegian Institute of Bioeconomy Research (NIBIO))

Municipality	Number of sheep and lambs in OBB 2017	Number of sheep and lambs in PT 2017	Difference
Hemsedal	4841	4504	337
Ål	21706	19333	2373
Hol	16614	12393	4221
Eidfjord	6940	1520	5420
Ulvik	6061	5970	91
Aurland	5147	5855	-708
Lærdal	9835	9925	-90
Sum:	71144	59500	11644

In addition to sheep, 1342 cattle and 3462 goats were sent on pasture in the NF-MA area in 2017. The bulk of the cattle grazed in the areas of Ål beitelag and Lærdalen sankelag, while goats were found in Ål, Vestfjelli, Hol and Lærdal.

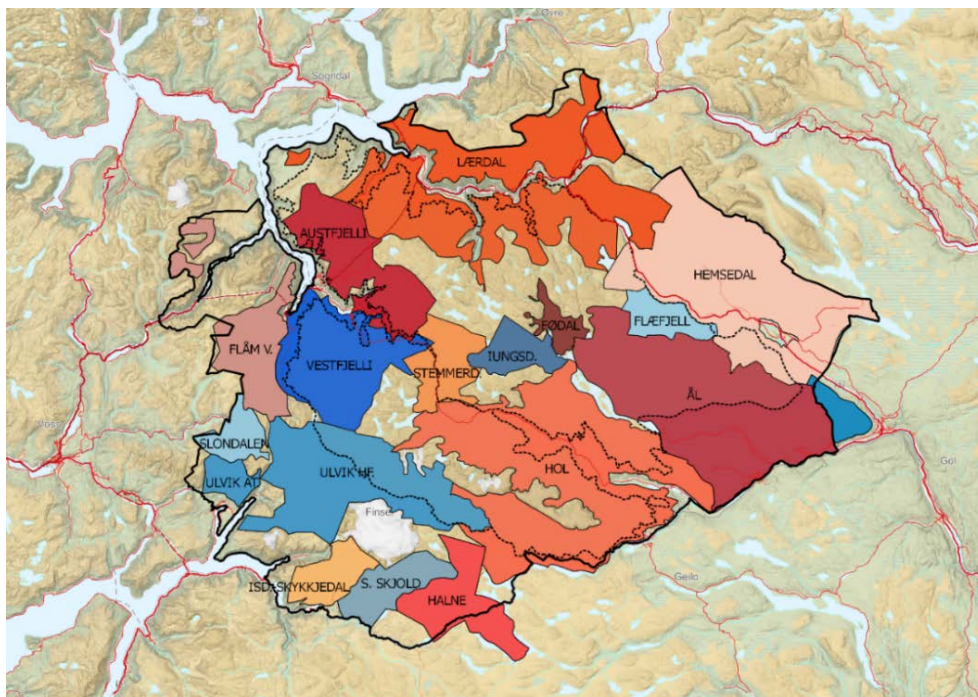


Figure 3-10. The areas utilized by beite-/sankelag for domestic animals in the NF-MA, defined with a black line. Nordfjella Wild Reindeer Area and the border between Zone 1 and 2 is defined with the dotted black line. Several of the beite-/sankelag utilize areas in several zones of the management area. (Data source: Organisert beitebruk/NIBIO)

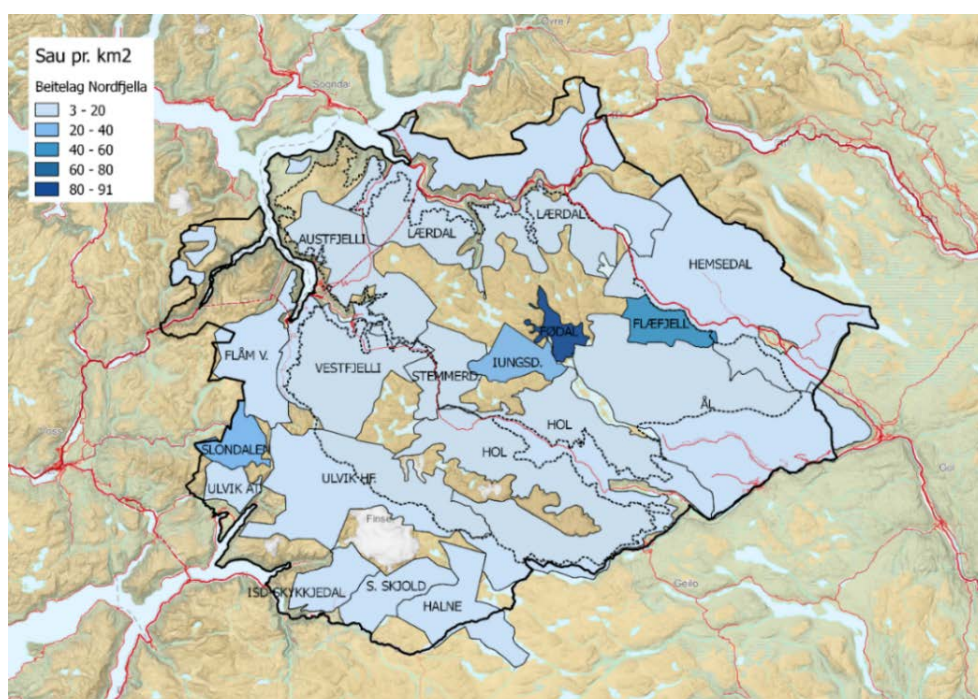


Figure 3-11. The density of sheep on pasture vary a lot between the beite-/sankelag areas, from 4 and 6 per square km in Flåm Vestfjell and Vestfjelli, to 37, 49 and 90 in Lungsdalen, Flæfjellsdrifta and Fødalsdrifta, respectively, in the central/eastern part of Zone 1. (Data source: Organisert beitebruk/NIBIO)

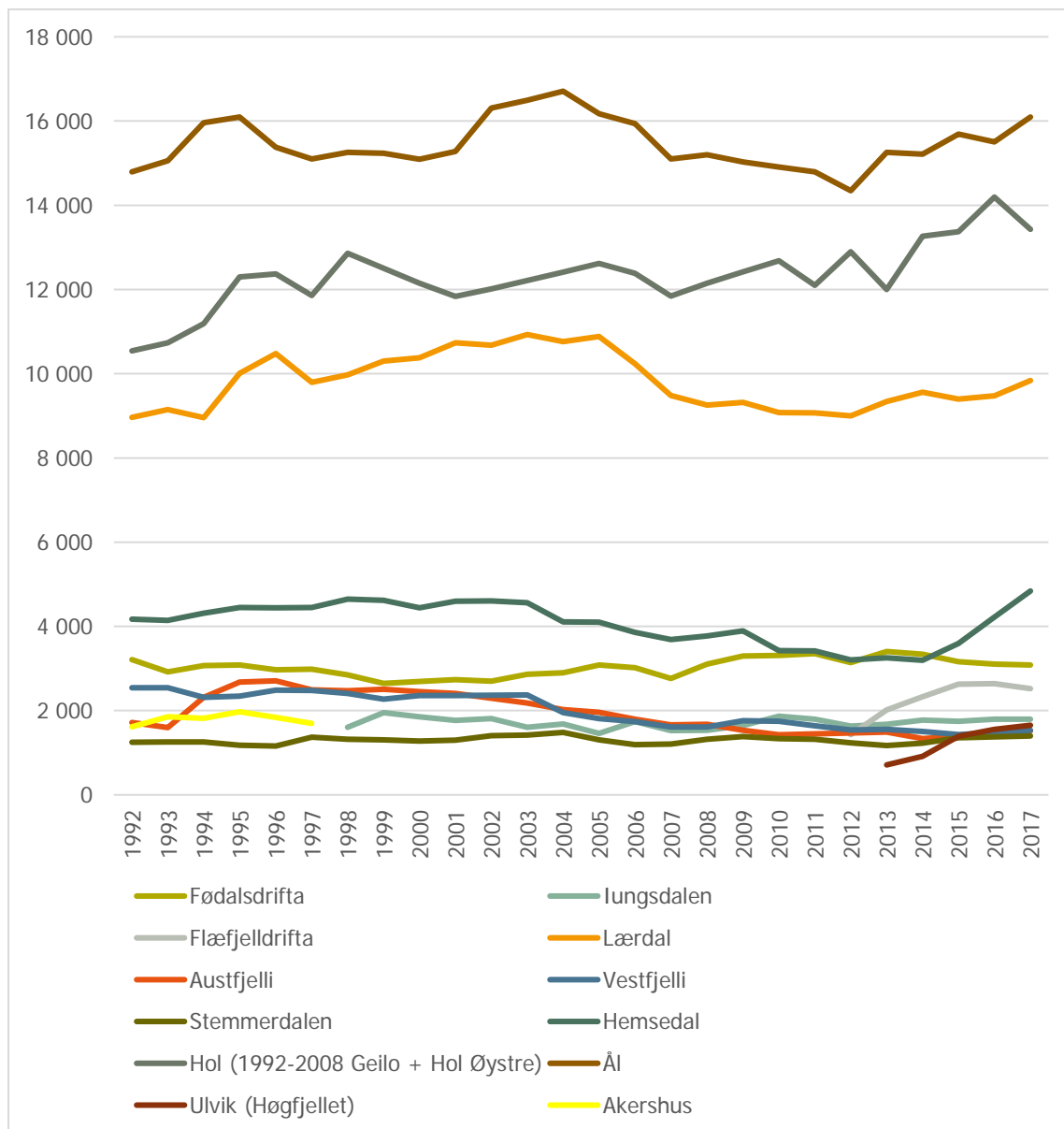


Figure 3-12. Number of sheep (ewes and lamb) released on pasture in the different beite-/sankelag at least partly using areas in the Nordfjella wild reindeer area, from 1992 to 2017. Data from Isdal-Skykkjedal, Søre Skjold and Halne beitelag in Eidfjord utilizing the pastures on Hardangervidda right north of RV 7 and Slondalen, Ulvik aust for Tysso and Flåm Vestfjell beitelag west of the railroad Flåmsbana, all in Zone 0, are not shown. (Data source: Organisert beitebruk/NIBIO.)

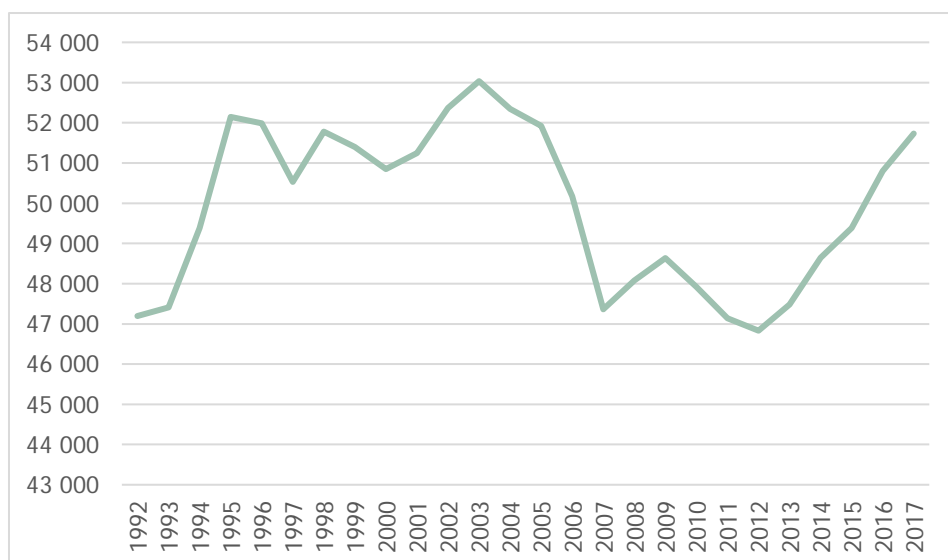


Figure 3-13. Development from 1992 to 2017 in the number of sheep (ewes and lamb) released by the beite-/sankelag Fødalsdrifta, Lærdal, Austfjelli, Vestfjelli, Stemmerdal, Hol og Ål utilizing areas that at least partially are located within NF-Z1. These represent app. 80% of the total amount of sheep currently released by beite-/sankelag in the entire NF-MA and has participated in organisert beitebruk the whole period. The total number of sheep released in the Management Area in 2017 was 65.612. (Data source: Organisert beitebruk/NIBIO.)

3.6 Use of mineral licks

There are to our knowledge no publicly available statistics for use of salt licks for sheep or sale of such. Mattilsynet was given access to sale statistics from 2010 to 2016 from GC Rieber, a company that according to their own web pages (<http://www.gcrieber-salt.no/markeder/landbruk/> accessed the 1st of October 2018) is the leading supplier of salt for all agricultural purposes. The figures of nationwide sale during this period increased from 663.572 kg in 2010 to 914.656 kg in 2016, i.e. an increase of 37.8% (Figure 3-14).

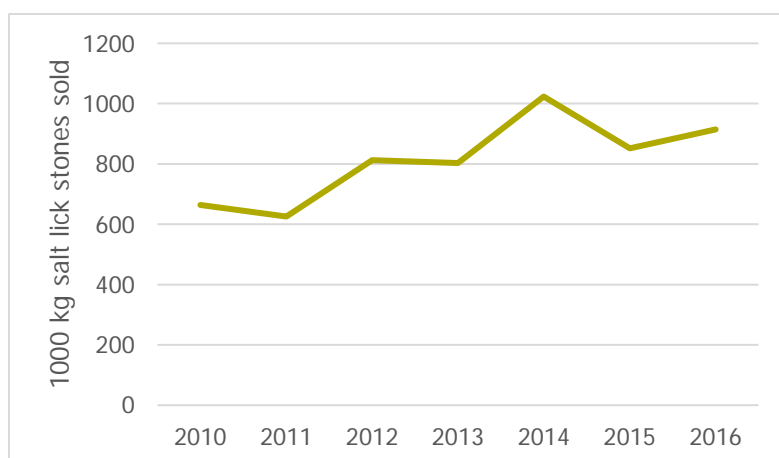


Figure 3-14. Sale of salt lick stones (tonnes) for husbandry use (all species) in Norway in the period from 2010 to 2016 from one supplier (Data source: GC Rieber Salt).

To our knowledge, there is only anecdotal information about how mineral licks have been used and the extent of the use in a longer time frame. Our perception is that in most areas, mineral licks were used on a relatively low number of places and mainly close to shielings or other buildings on mountain pastures or on places where sheep were gathered in autumn. Our impression is that it used to be more common to spread smaller amounts of feed salt or use single lickstones only three to four decades ago, while today, lickstones are used more systematically to improve and ease the herding of sheep on rangeland and limit their dispersal outside grazing areas, facilitating optimal use of pasture resources. Some sheep farmers express that increased herd sizes (see above) and an increased proportion of owners having sheep farming only as a part of their income, necessitates arrangements that allow easy and time-efficient management of the animals. However, these perceptions are difficult to document and there is a lack of knowledge of the individual farmer's motivation for his/her use of lickstones and the societal drivers responsible for the development. Based on the map describing the locations of the mineral licks, how they are used also seem to vary between the individual sanke-/beitelag areas (Figure 3-15).

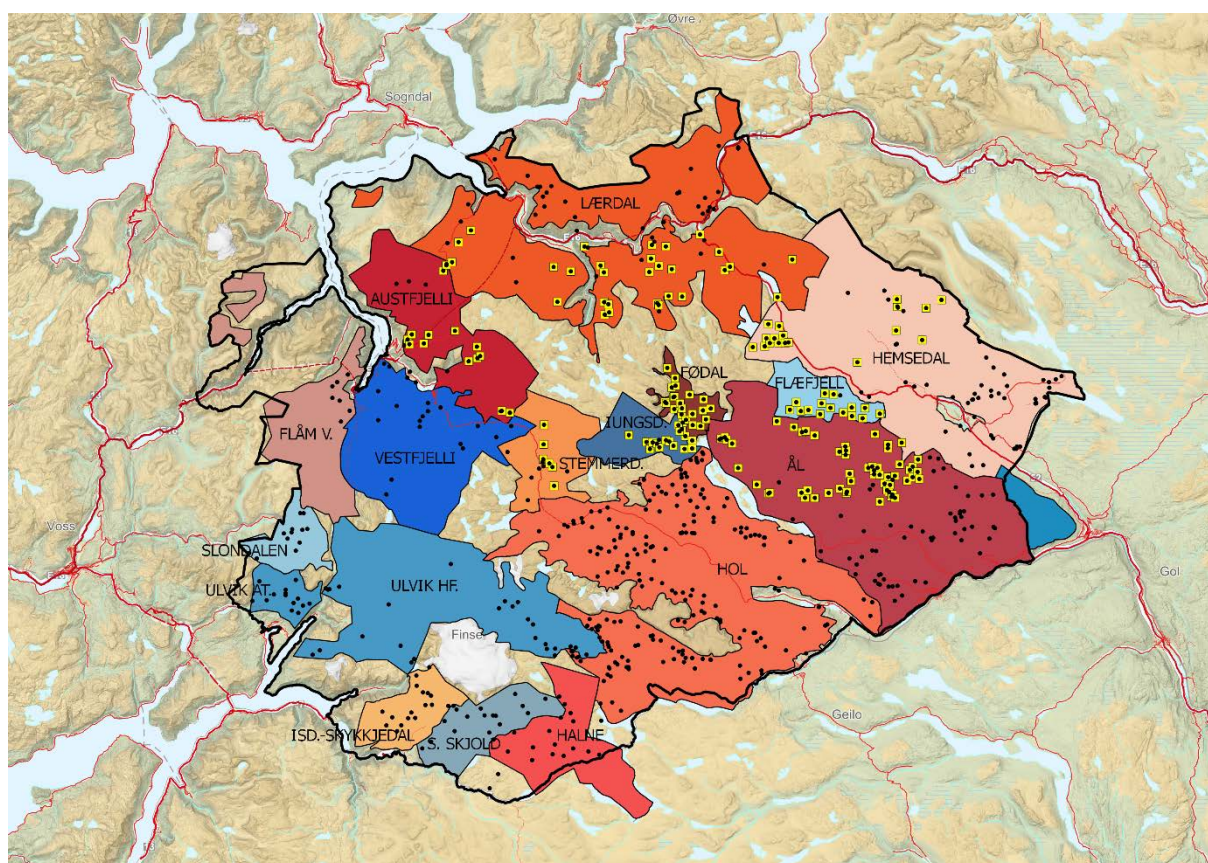


Figure 3-15. Positions of mineral licks that are completely fenced off (black dots surrounded by yellow square) and mineral licks with openings designed to allow access for sheep (black dots) in the different beite-/sankelags throughout the NF-MA. Based on this map, the mode of use of mineral licks seem to vary between different beite-/sankelag areas, but little is written about how mineral licks are used for sheep and pasture management. (Data sources: Organisert beitebruk/NIBIO and NFSA)

3.6.1 Mineral licks and animal movements

There is a lack of quantitative data on the role of natural and artificial salt licks for cervids' space use in Scandinavian ecosystems. Soil ingestion at natural mineral licks (Kreulen 1985; Klaus & Schmid 1999), use of road salt (Tiwari & Rachlin 2018), antler consumption (Gambín et al. 2017) and even predation (Bazely 1989) by cervids presumed to be motivated by a need for minerals, are well-known phenomena. The cervids' propensity to consume minerals according to nutritional requirements is known from experimental studies of red deer in Spain (Ceacero et al. 2010). Studies of species like white-tailed deer (Atwood & Weeks 2002), sika deer (Ping et al. 2011), and moose (Tankersley & Gasaway 1983) show seasonal variation in use of salt licks and patterns of use differ among males and females. Mineral composition of natural salt licks can benefit mineral intake (Ayotte et al. 2006), and they were frequently used during summer by wapiti and moose in Canada (Ayotte, Parker & Gillingham 2008). Licks are mainly used from spring to fall, and particularly in summer when body growth and reproductive costs are at the highest. In Canada, moose may use salt pools during spring and summer in areas along highways (Laurian et al. 2008). Anecdotal observations in Norway suggest that cervids visit roads to lick salt during autumn and winter when road agencies spread salt on the roads to avoid icing (C.M. Rolandsen, pers. comm.).

Salts are generally not regarded as a limiting resource for cervid populations in Norway. However, given that artificial salt licks are available within their summer home range, we find it likely that this may affect the space used by of animals. During summer, boundaries of cervid home ranges are partly determined by social factors and kinship. Movements outside of ordinary home ranges are unusual. Thus, it is considered less likely that salt licks influence the choice of summer home range and its boundaries.

It is therefore difficult to assess whether the artificial mineral licks for sheep in Nordfjella attract additional numbers of cervids to the region without further quantitative studies of cervids use of mineral licks.

Red deer and moose will predominantly use salt licks positioned in the forested areas. During night-time, however, both red deer and moose visit alpine areas above the treeline to some extent and are likely use mineral licks a few kilometres above the treeline.

3.7 The difficulty of restoring mineral licks

Methods that efficiently inactivate environmental CWD contamination are, to the best of our knowledge, not available. A recent report from Kuznetsova and co-workers (Kuznetsova et al., 2018) demonstrate that humic acids that are naturally present in some soils can significantly degrade and inactivate CWD prions. These results are encouraging and merits further studies.

Anecdotal information with some support in the literature (Schramm et al., 2006) suggest that salt-laden soil surrounding mineral licks is attractive for deer for many years after the removal of the salt stone itself.

When classical scrapie is diagnosed in sheep herds, or CWD in farmed deer, strong disinfectants are used wherever suitable, surfaces are removed or painted and soil etc. from areas frequently used by the infected herd is either removed or covered.

Most of the mineral licks in Nordfjella are now closed by fences. However, many of the fences are equipped with openings that many cervids go through, apparently without effort (Figure 3-16). Moreover, the fences normally cover only part of the area that receive run off from the mineral stone. Therefore, areas surrounding the fences might be attractive to deer. The size of this periphery will vary significantly between sites depending upon vegetation and soil. For instance, urine and faeces from the visiting animals will fertilize the area and stimulate plant growth outside the eroded area, making the spot an attractive pasture (Figure 3-17). We do not have methods that reliably allow detection of environmental CWD prions to ascertain which mineral licks that are contaminated and which (and how large) areas of the mineral licks that contain infectious material available to susceptible animals.



Figure 3-16. Wild reindeer in NF-Z2 visiting a fenced mineral lick. An animal is passing through the opening on the left side of the fence, while another is licking on the mineral stone and a third one seem to find something palatable on the ground outside the fence. (Photo: NINA Wildlife Camera)



Figure 3-17. A mineral lick in Forolhogna Mountains is visited by a small herd of wild reindeer. The animals have dispersed out in the eroded area and seem to lick or eat soil. Around the eroded area, the vegetation is considerably greener than in the surrounding areas, presumably due to the fertilizing effect of urine and faeces from the animals. In a situation where there is an outbreak of CWD, we do not know where and for how long time CWD prions are accessible for susceptible animals.

3.8 Human activities in Nordfjella

Nordfjella is widely used for recreation, fishing and hunting. Area usage patterns and intensity has been studied and reported by Strand et al. (2011) and Gundersen et al. (2013). As in other mountain areas of Norway, the local inhabitants utilize the mountains in a relatively traditional way as pasture for their livestock, and for harvesting (berry-picking, hunting and fishing). In addition to the locals, visitors also use the area for fishing and hunting. These harvesting activities are typically dispersed over large parts of the area.

Nordfjella also have a high number of private cabins in addition to several hotels and tourist resorts. In total, there is approximately 13 200 private cabins in the area, mostly on the eastern fringes of the mountain range. Commercial tourist enterprises in the area report approximately 1.4 million guests annually, but only a minor and unknown proportion of these guests venture into the mountain areas. Most visitors trek along marked trails and hiking routes from relatively few entrance and exit points. The visitors' use of the area varies spatially, being more intense in Zone 2, and, in particular, along the road Rallarvegen and marked trails like the route between the tourist cabins at Finse and Geitryggen (Figure 3-17). This is also the area with highest overlap between wild reindeer and human visits. Usage

patterns and intensity also varies on a temporal basis. In general, winter use is less intense, but still relatively high in the eastern parts of the area, where a high number of cross-country skiing trails crisscross the area.

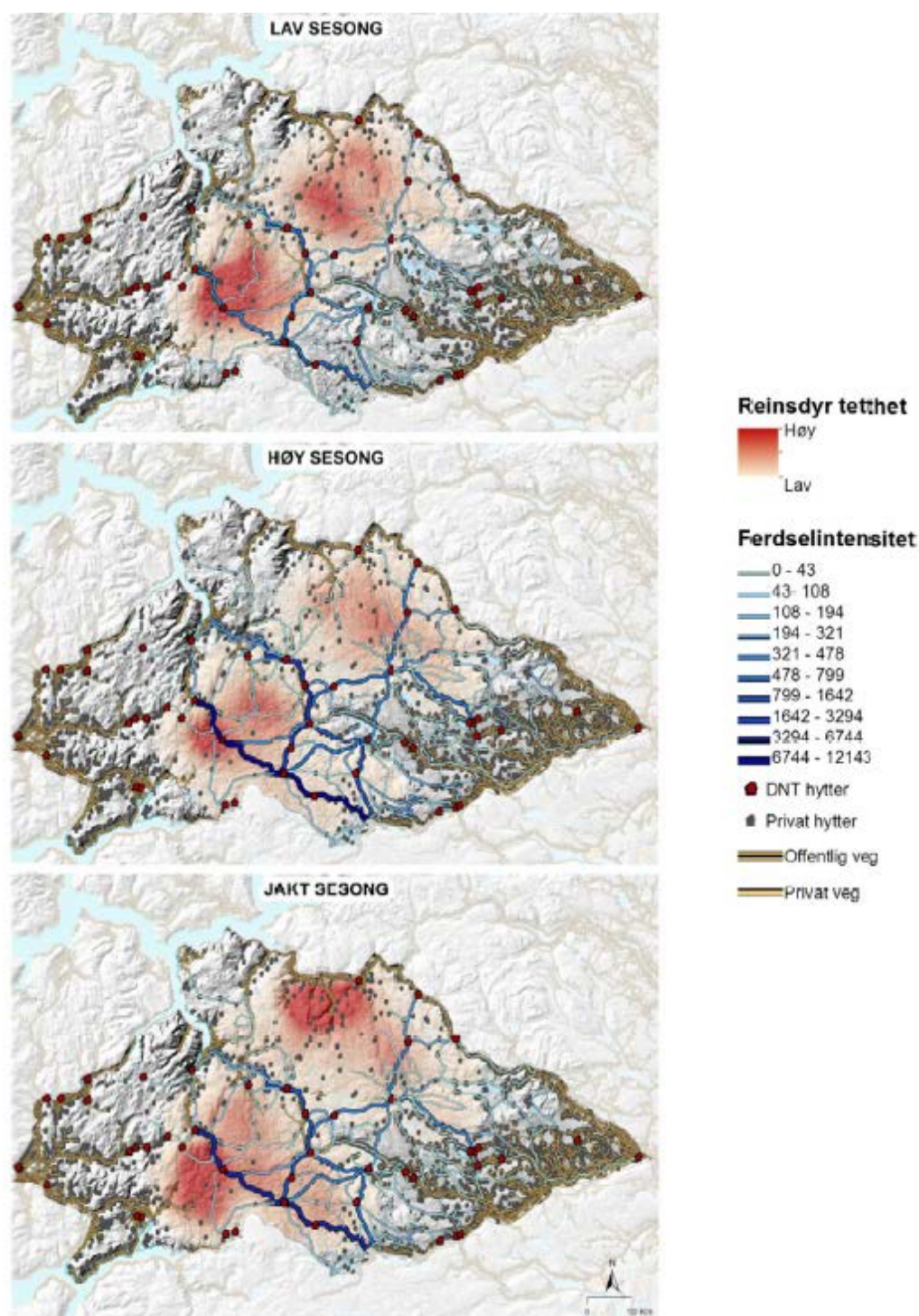


Figure 3-18. Map of Nordfjella with estimates of reindeer density areas (in red) from GPS data and the intensity of tourist trail use (lines in different colours and thickness) based upon data from trail counters. “Lavsosong” means low season, “høysesong” high season (i.e. summer) and “jaktosong” hunting season (Source: (Gundersen et al., 2013))

4 Exposure characterisation

4.1 Environmental contamination in Nordfjella

Among the approximately 2400 wild reindeers from Nordfjella Zone 1, that were examined with conventional methods (i.e. Enzyme Linked Immunosorbent Assay test for detection of abnormal prion protein (TeSeE ELISA Bio-Rad), nineteen tested positive for CWD. Further detailed investigation of the positive animals showed that in 9 (47 %) of these, only the retropharyngeal lymph node was positive, indicating that they were in an early phase of the infection. In the remaining ten, also the samples from the brain stem turned out positive, but immunohistochemistry and histopathology showed that neither of these cases were in an advanced stage of the disease (Pers. comm. S. Benestad), and apart from the first case, none of the animals showed clinical signs of disease. In addition to these nineteen cases, we assume that there have been cases of diseased animals that not have been discovered and examined but died of the disease or related causes. An example of such a missed case was a male that highly unusually attacked one of the reindeer herders that guarded the border between Zone 1 and Filefjell in January 2017, i.e. close to RV52. The buck was salivating, seemed emaciated and did constantly eat snow - clinical signs that could be consistent with CWD (Figure 4-1).



Figure 4-1. Lone wild reindeer male in Nordfjella Zone 1 in January 2017 showing signs that could be consistent with CWD, such as abnormal behaviour (aggression), heavy salivation (reduced ability to swallow), constantly eating snow, presumably due to dehydration, frequent urination, and loss of shyness. The reindeer was searched after later, but was not found (Photo: Runar S. Bjøberg)

In addition, it is reasonable to assume that there has been an unknown number of undiagnosed cases in the early phases of infection among those examined, since there is uncertainty about the diagnostic sensitivity of ELISA tests (Ricci et al., 2018). The estimated apparent (i.e. detected by ELISA) prevalence among adult reindeer in Nordfjella was 1.2%, but the true prevalence was estimated at 1.6% after accounting for imperfect detectability (Viljugrein et al., 2018). As expected, sensitive amplification methods, such as protein misfolding cyclic amplification (PMCA) or real-time quaking-induced conversion (rtQuIC) diagnose more cases than conventional methods when used on the same sample of individuals (Haley, Seelig, et al., 2009; Selariu et al., 2015).

As all reindeer use large parts of the available area, in contrast to other cervids who typically stay in a more limited home range, a large proportion of Zone 1 must be regarded as exposed to contamination from infected animals. This is exemplified by the wide distribution of places where the positive animals were shot or found (Figure 4-2) and by the maps illustrating the area use of reindeer (Figure 4-4 and Figure 4-5).

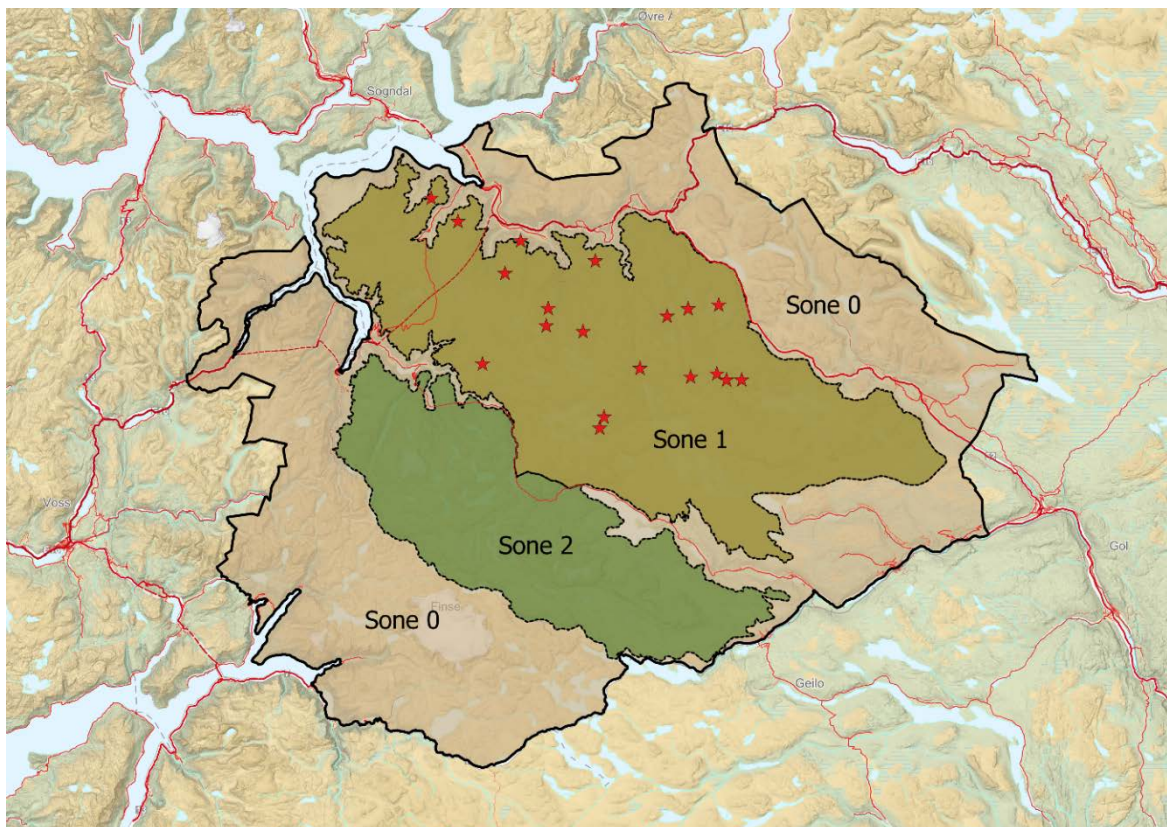


Figure 4-2. Map describing the positions where the nineteen infected animals were found/shot. Notably, they are dispersed over a large proportion of Zone 1, illustrating that an equally large area should be considered exposed to contamination with CWD.

In estimating/evaluating the overall environmental CWD contamination in NF-Z1, the following assumptions were made:

1. Low number of infected animals, dispersed over a large area
2. Relatively early stage of outbreak
3. High precipitation levels – heavy rain- and snow-falls, wash-out effects
4. Low levels of clay in soils
5. Spring and autumn periods with repeated freezing/thawing cycles, possibly reducing prion infectivity
6. High number of man-made mineral licks for sheep, creating aggregation points for livestock, reindeer, red deer and moose.

Preliminary observations in the study mentioned in chapter 1.3.5, indicate that mineral licks attract wild reindeer, which aggregate there and disperse out on the eroded ground surrounding the mineral stone itself, and that they lick and ingest soil there, presumably because it is loaded with salt run-off. A mineral lick consequently creates a spot which not only attract many animals over time, and where these animals aggregate (which alone increase the risk of disease transmission), but also a place where the animals are stimulated to ingest salty soil and vegetation potentially contaminated by other animals' excreta. Considering that prions can bind to soil as well as vegetation, and that this in some instances increases their infectivity (Johnson, Pedersen, Chappell, McKenzie, & Aiken, 2007; Wyckoff et al., 2016), man-made mineral licks and their surroundings must be considered significant hot-spots for transmission of CWD in any area this disease occurs.



Figure 4-3. The CWD positive wild reindeer no. 236 photographed while immobilized during collaring (left) and after (right). (Photos: Roy Andersen, NINA.)

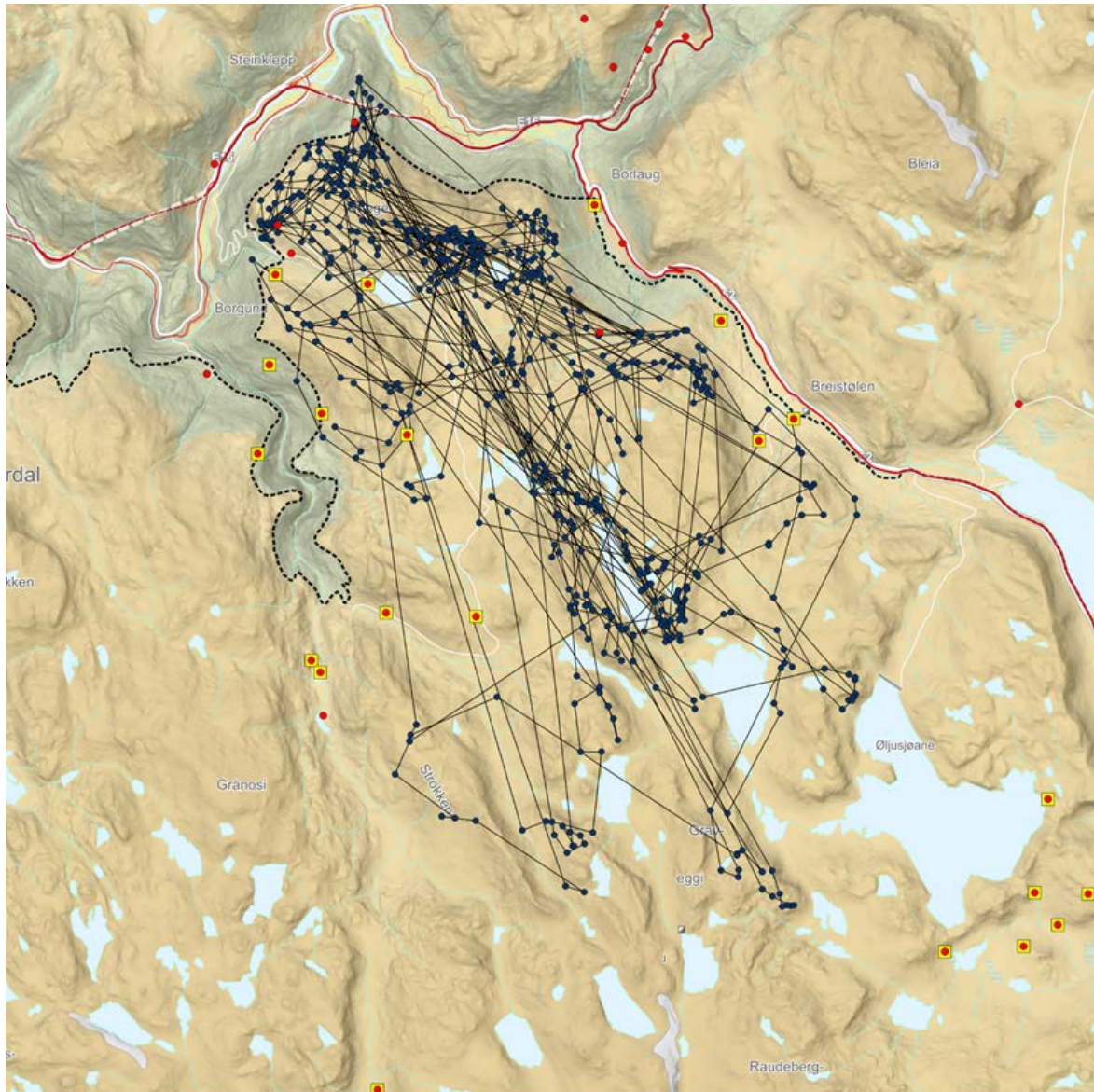


Figure 4-4. GPS-positions from wild reindeer no. 236 equipped with radiocollar 46. Biopsies taken from the rectoanal mucosa-associated lymphoid tissue at marking were diagnosed positive for CWD prions by immunohistochemical examination at the Veterinary Institute. The map shows its movement from the 29th of March, when it was collared, to the 21st of June 2017, when it was shot. Positions were registered every third hour if the collar was within coverage of the GSM network. Hence, the points and their connecting lines do not depict the detailed tracks of the reindeer. Still, it is evident that the reindeer has been very close to several mineral licks that are now fenced in (red dots in yellow squares) and mineral licks that are still accessible during the sheep grazing period (red dots). Based on observations from ongoing studies, it is likely that the animal visited mineral licks on several occasions.

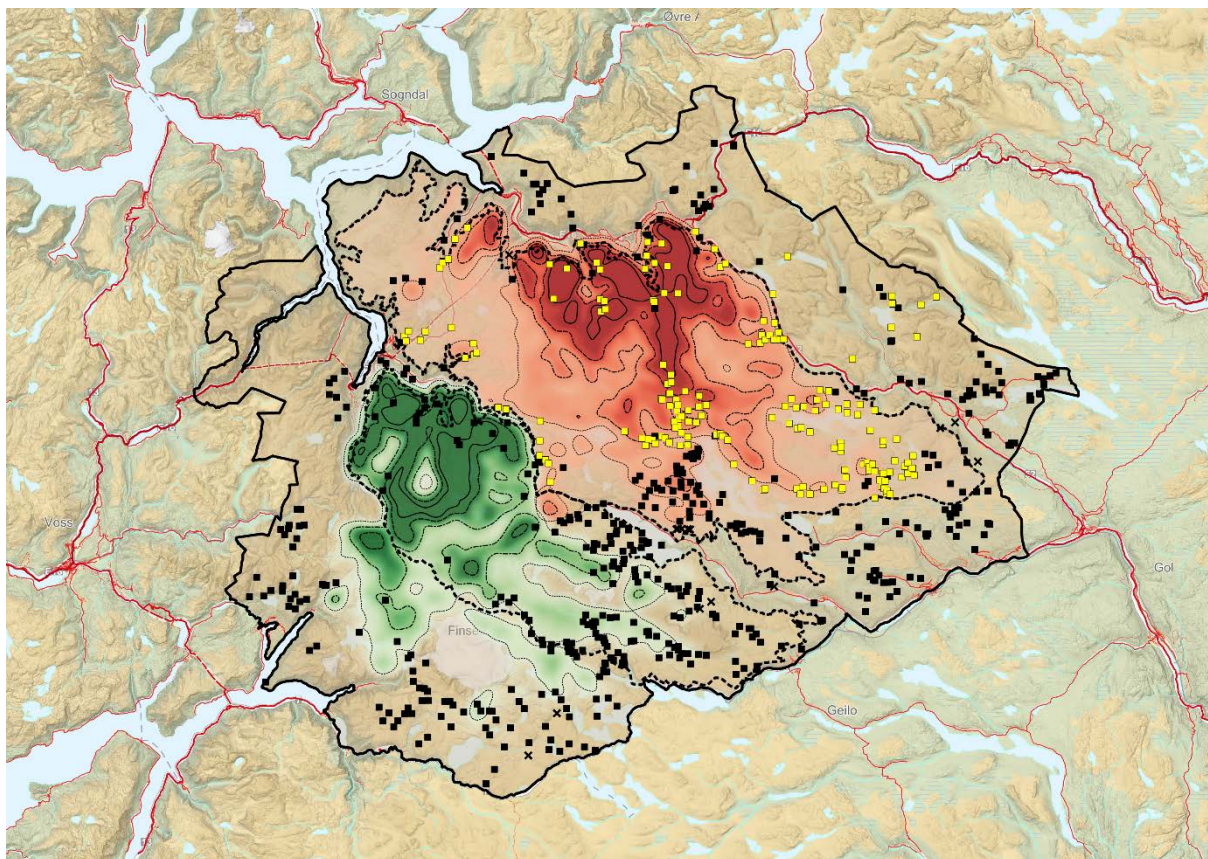


Figure 4-5. Map illustrating the area use of the reindeer herds of NF-Z1 (red) and NF-Z2 (green) based on GPS positions from 31 and 27, respectively, collared reindeer collected in the period from 2007 until now in relationship to the positions of the mineral licks. Relative density of wild reindeer as estimated from GPS fixes as 95, 75 and 50% kernels. Mineral licks are shown as black squares, while mineral licks that are registered as completely closed with fences are indicated with yellow squares. The darker red/green colour, the higher is the density of reindeer positions. NF-Z1 and NF-Z2 are demarcated with stippled lines. It is evident that many mineral licks are located within areas that frequently were used by reindeer from the infected herd. Notably, only 5 of 31 marked animals in NF-Z1 and none in NF-Z2 were males. Consequently, the map does not give a representative picture of the movement pattern of male reindeer in the area. (Positions of mineral licks were supplied by NFSA. Map source: Geir Rune Rauset and Olav Strand, NINA)

4.2 Transmission of CWD to reindeer

Influx and/or transit of reindeer into NF-Z1 is considered to increase the probability of spreading the prions throughout the fallow period, although the level of environmental CWD contamination is expected to decrease significantly from one year to another. Reindeer are considered highly susceptible to the CWD strain in question. Therefore, the probability of a reindeer getting infected is suspected to be substantially higher than any other animal, with the possible exception of other cervids (see discussion below). Therefore, effective measures to minimize reindeer influx into NF-Z1 should be evaluated.

Since we know that there has been contact between the infected herd in NF-Z1 and the reindeer herds in NF-Z2 and with Filefjell reinlag, and that animals, at least from Filefjell, also after the eradication have visited NF-Z1, we cannot exclude the possibility that infected reindeer already are present in these populations. In Filefjell reinlag, a significant number of animals (about 900) have been tested since 2016. In addition, several preventive measures are instated and, importantly, the herd is closely supervised during herding, so that animals with clinical signs of disease are more easily identified and examined. In contrast, relatively few animals have been sampled from NF-Z2, and the herd is difficult to survey due to the rough terrain and unstable weather in their area.

As described above (chapter 3.4.1.3), reindeer have been translocated from Nordfjella on several occasions. The low prevalence indicate that the outbreak was discovered in an early phase. Based on comparison with outbreaks in North-America (though they have occurred in different cervid species and with different strains) it is reasonable to presume that the prion disease not has been present in Nordfjella for many decades. At the same time, however, it appears plausible that CWD became established in Norway more than a decade ago. (Ricci et al., 2018). In VKM report 2017:9 we suggested that the disease first occurred up to 15 years ago. Consequently, there is substantial uncertainty about when CWD first occurred in Nordfjella, and we cannot completely exclude that any of the described translocations of reindeer included infected animals.

However, it is less likely that the translocations that occurred thirty – forty years ago to Sweden and other places, have caused spread of disease, than more recent contacts between NF-Z1 and susceptible animals. The transport of wild reindeer to Lærdal-Årdalsfjella in 1994 – 1995 can also have occurred before CWD was established in NF-Z1, but as we do not know the transmission rate of the present strain in reindeer, we cannot exclude the possibility that infected animals were translocated, and intense surveillance of that population is warranted. Among the described translocations, the greatest uncertainty and highest probability of transmission is related to the semidomesticated reindeer kept on Geitrygghytta until 2016. The tunnel nearby is a well-known crossing point between NF-Z1 and NF-Z2, and it is most probable that these semidomesticated animals roamed around in both areas. We do not know when or where animals were transported from this herd to other herds in Scandinavia, but anecdotal information indicate that this has occurred, also relatively recently.

4.3 Transmission to other cervids (preclinical and/or nonclinical carriers)

We do not know to what extent the CWD strain observed in Nordfjella is transmissible to species other than reindeer, but precautionary principles suggest that all deer species should be considered highly susceptible until proven less susceptible. Likewise, it is assumed that this CWD strain, as those studied in North America, has a very low transmissibility to other non-cervid species such as sheep (Hamir et al., 2006), cattle (Hamir, Miller, Kunkle, Hall, & Richt, 2007) and, importantly, humans (Kurt et al., 2015).

Thus, although susceptibility to CWD can vary among deer and be subject to host *PRNP* genetic modulation (variable incubation periods) (Brandt et al., 2015; Duque Velasquez et al., 2015; Johnson et al., 2011; Robinson, Samuel, O'Rourke, & Johnson, 2012; Wilson et al., 2009), it is likely that the infective dose necessary to establish (and amplify) the agent in a deer species is substantially lower than for any non-cervid. Therefore, the probability of exposed deer becoming preclinical carriers is considered much higher than for instance cattle or sheep (see below), when being exposed to similar levels of infectivity.

The combination of relatively high susceptibility (as assumed for deer species), with very low levels of environmental prion contamination can, at least in theory, generate preclinical carriers with abnormally long incubation periods. This calls for continued comprehensive surveillance of all deer populations surrounding NF-Z1.

Based upon data on habitat overlap between wild reindeer, red deer and moose, and the localization of man-deposited mineral licks, it is evident that spill-over of CWD from contaminated environs in Nordfjella to red deer in the Northern parts of NF-Z1 is more likely than to moose.

If red deer are as susceptible to the Nordfjella prion strain as reindeer, the level of influx and transit of red deer into certain areas of NF-Z1 might constitute a significant potential for dissemination of the disease. Also, CWD would be much harder to control in red deer populations that are more open compared to reindeer populations.

4.4 Transmission by sheep and other livestock (nonclinical and/or passive carriers)

Considering the current situation in Nordfjella, with high numbers of sheep grazing within Zone 1, the question of whether sheep can spread CWD prions, was noted as an important "knowledge gap" in the Phase 2 report by VKM (VKM, 2017b). As mentioned, investigations to address this have been initiated (see 1.3). The primary objectives are to clarify whether sheep:

- a) are susceptible to the Nordfjella CWD strain, that is; whether they develop clinical prion disease and thus theoretically can operate as a "preclinical carriers"
- b) can take up and replicate this CWD strain silently and function as a nonclinical carrier

It is also likely that neither "a" nor "b" will be the case, which would suggest that sheep should be grouped together with other animals in the "Passive carrier" category (see below).

It should, however, be kept in mind that even if the probability of sheep getting infected by CWD should be extremely low, the high number of sheep visiting potentially contaminated environments in NF-Z1, raises the overall probability of transmission to levels that must be taken into consideration.

4.5 Transmission with passive carriers

Studies performed in rodents (Kruger et al., 2009; Maluquer de Motes et al., 2008), coyotes (Nichols, Fischer, Spraker, Kong, & VerCauteren, 2015) and crows (VerCauteren, Pilon, Nash, Phillips, & Fischer, 2012) have shown that ingested prions can passage through the gastrointestinal tract and still retain infectivity. An *in vitro* experiment furthermore indicated that prion infectivity is not diminished after ruminal digestion (Samuel E. Saunders, Bartelt-Hunt, & Bartz, 2012).

This evidence indicates that animals that actually are exposed to environmental prions, are likely to carry them, associated with soil and dust in their pelage, on their feet and in their gastrointestinal tract, in the content of the gastrointestinal tract or bound to plastic or metals in earmarks, collars or bells, and that prions released from the animals exterior, their faeces or man-made objects they carry may be infective to susceptible animals.

However, the true epidemiological impact of this cannot be estimated because we lack knowledge about:

1. The level of CWD infectivity in environs in Nordfjella Z1
2. To what extent grazing animals are exposed to contaminated environs
3. If exposed, which quantities of prions can be carried and later released
4. What are the infective doses of this CWD strain needed to transmit the disease to a susceptible host – the levels of infectivity released by a passive carrier are considered to be very low.
5. What is the range of susceptible hosts for the CWD strain?

Concerning points 1 and 2 above, the man-made mineral licks that are placed in areas with significant overlap between wild reindeer, red deer and sheep, constitute the most important elements that should be looked at for minimizing the risk of spread of CWD prions from contaminated environs in Nordfjella Z1.

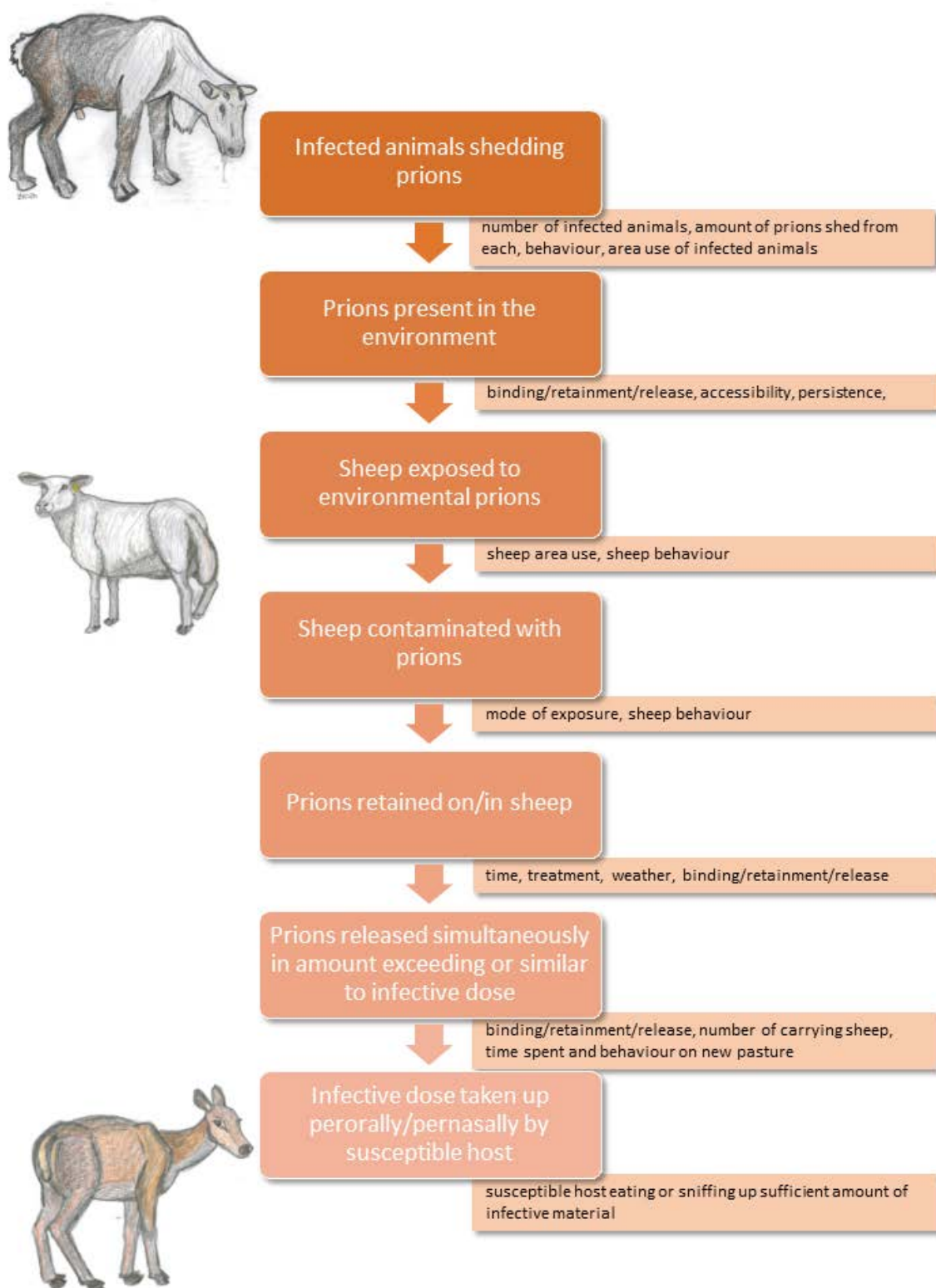


Figure 4-6. There are several critical steps, each influenced by many factors, that are necessary if mechanical/passive transmission from reindeer in Nordfjella via the environment and sheep to susceptible cervids in other areas should occur.

4.6 Mechanical transport of CWD infectivity

Likewise, Nordfjella and the surrounding mountains are popular tourist destinations. Trails zigzag the area and hundreds of privately owned mountain cabins are scattered in the lower areas and in the periphery of the mountain plateau. The extent to which human recreational activity in the NF-Z1 and its surroundings constitute a risk of passive dissemination of CWD infected materials will be discussed.

4.7 Concluding remarks on exposure characterisation

According to current knowledge, the following reservoirs have the capacity to hold infective doses of prions and can hence function as a source of continued spread of CWD.

Importantly, we mostly consider environmental point-sources to be epidemiologically relevant in Nordfjella. We do not expect the whole area to be uniformly contaminated with CWD prions. Identification of areas that are likely to be point-sources of CWD contamination is therefore crucial:

1. soil, water, plants and surfaces (rocks, man-made materials) at mineral licks used by the now eradicated wild reindeer population of Zone 1 – have been identified
2. eventual remaining carcasses of infected animals in Zone 1 and the soil underneath them – unlikely to be identified
3. other contaminated areas in Zone 1 – unlikely to be identified
4. potential red deer within the Management Area that have been infected by direct/indirect contact with the now eradicated wild reindeer herd of Zone 1
5. potential infected wild reindeer in Zone 2 (or other recent contact populations)
6. potential infected semidomestic reindeer in Filefjell reinlag
7. potential infected moose or roe deer in the Management Area
8. equipment, vehicles or garment contaminated in Nordfjella Zone 1 before or after the eradication of the herd
9. soil on home pastures contaminated by prion-containing dirt transported with passive carriers (sheep that grazed in Zone 1) and potential cervids in these localities that have been infected due to contact with this
10. potential nonclinical carriers among sheep
11. passive carriers among other animals (cattle, goats, rodents, shrews, scavengers (like crows, ravens, eagles, foxes, wolverines etc.), insects, dogs, humans)
12. potential non-clinical carriers among other animals (cattle, goats, rodents, shrews, scavengers (like crows, ravens, eagles, foxes, wolverines etc.), insects, dogs, humans)

5 Conclusion with answers to the request

This report aims to provide an answer to the request to provide information about factors that can contribute to the spread of Chronic Wasting Disease (CWD) within and beyond the Nordfjella-area.

As illustrated in the Figure 5-1, the risk of spillover of CWD from contaminated environs in NF-Z1, can be schematically presented according to these classes of “activities”: Reindeer, Other Cervids, “Livestock (sheep)” and “Human (recreational)”.

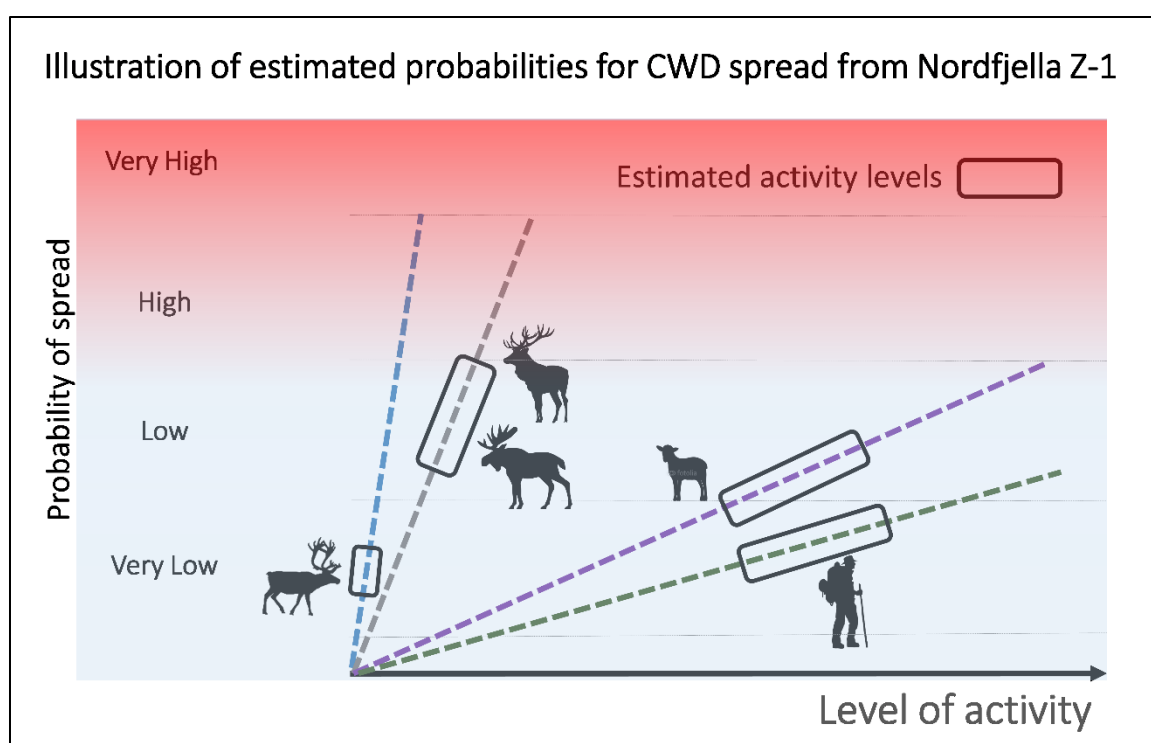


Figure 5-1. Conceptual and schematic diagram illustrating probability of CWD spread from NF-Z1 (Y-axis) given certain levels of activities (X-axis). The steepness of the dotted lines illustrate how easily an animal or human can contribute to spread of CWD. The steep line for reindeer shows that we consider reindeer to be highly susceptible to this CWD strain and that only a modest increase in activity (i.e. reindeer crossing into NF-Z1) cause a rapid increase in probability for spread.

We have, in lack of data grouped moose and red deer together on a line somewhat less steep than reindeer, assuming a little lower susceptibility. Note that red deer is localized higher on the curve than moose, reflecting their considerable habitat overlap with reindeer in some areas of NF-Z1. Sheep and other livestock are considered to be far less susceptible to CWD, although this is not known for this CWD isolate. The high number of sheep grazing in areas overlapping with reindeer makes this activity important to consider and is therefore

grouped at the same level at the Y-axis (risk of spillover) as red deer in this diagram. Human activity is considered to be of lesser importance for the spread of CWD, even if this activity occurs at a high level. The actual probabilities of CWD spillover are not known. The figure solely serves illustration purposes. Whether the relationship between “levels of activity” and “probabilities of CWD spillover” is linear or not, is not known. In the figure, straight dotted lines are used for simplicity.

From the above figure, it is evident that we consider red deer and sheep grazing in NF-Z1 to be the two most important activities with regard to possible spread of CWD prions. The figure also illustrates that activities of reindeer in NF-Z1 must be monitored closely and kept at a minimum. Given that point-sources, like mineral licks, are considered to be important sites for possible CWD dissemination, it is important to note that efforts that target and, if possible, eliminate such sites, can reduce the overall chances of disease spillover significantly. In Figure 5-2, the today's estimated risk related to sheep grazing is compared with a situation where sheep are denied access to environmental “hotspots”. The drop in steepness of the curve illustrates the importance of keeping sheep away from possible point-sources that are likely to be prion contaminated. Manmade mineral licks in certain areas in NF-Z1 are such sites. Importantly, by addressing this, the risks associated with red deer also automatically drops, since many of these sites are localized in the areas most frequently visited by red deer.

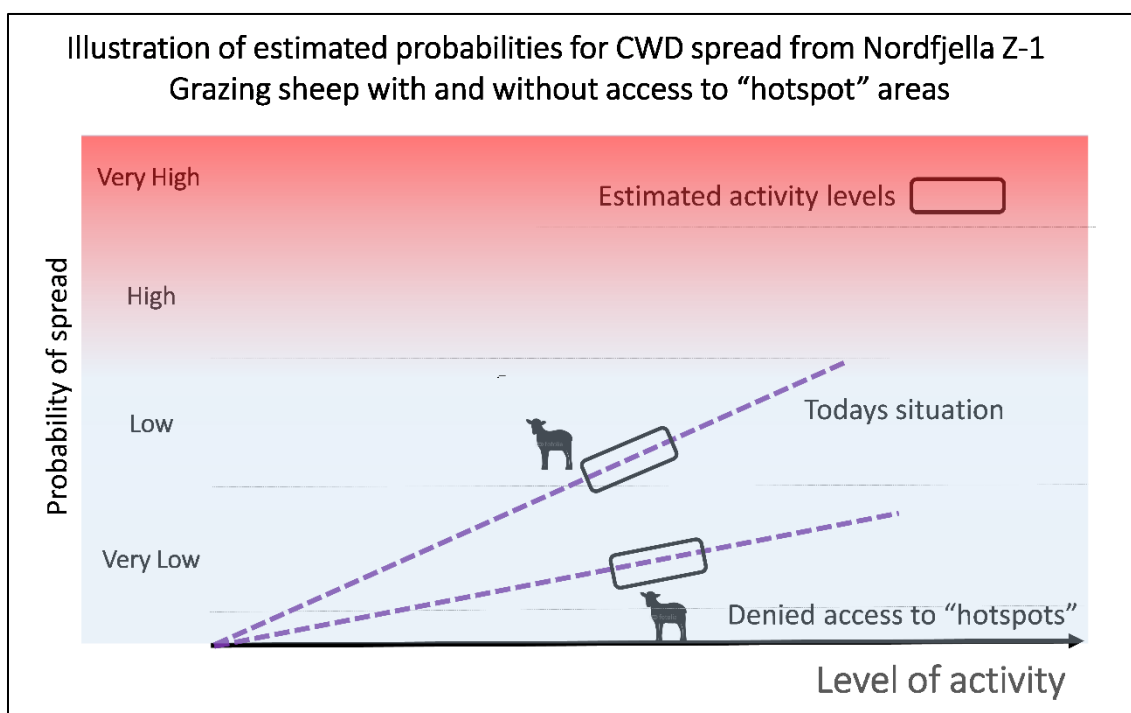


Figure 5-1. Conceptual and schematic illustration of reduction in probability of CWD spread related to sheep grazing when sheep are denied access to environmental hotspots that are believed to be contaminated with CWD prions. The level of activity is similar, but the probability of CWD spillover is reduced, as illustrated by the different slope of the two curves. The actual probabilities of CWD spillover are not known. The figure solely serves illustration purposes. Whether the relationship

between “levels of activity” and “probabilities of CWD spillover” is linear or not, is not known. As in figure 5-1 straight dotted lines are used for simplicity.

Below, we summarize the main elements discussed in this report, regarding probabilities for dissemination of CWD prions within and beyond Nordfjella. The emphasis is on variables that can be adjusted through management:

- Continued use and/or inadequate closing/clean-up of mineral licks previously used by wild reindeer in Zone 1
 - High probability of CWD transmission. Wild reindeer from Zone 2 or semi-domestic reindeer from Filefjell reinlag entering Zone 1 and visiting mineral lick areas.
 - Moderate to high probability of transmission. Entry of red deer and moose into Zone 1 with visit of mineral lick areas. We do not know the sensitivity of red deer or moose to the Nordfjella CWD strain, but high sensitivity is assumed. High-density populations of red deer and moose probably increase the number of animals entering Zone 1.
 - Low to moderate probability for passive transmission. Sheep visiting mineral lick areas and subsequent passive spread to cervids
 - Low/unknown probability for spread via nonclinical carriers among sheep
 - Very low probability of transmission via sheep that are infected and develop clinical CWD

Taking into consideration that CWD-infected cervids might be present (undetected) in contact populations surrounding Zone 1 (i.e. Zone 2, Filefjell reinlag, red deer, roe deer and moose in the Management Area), the following scenarios are relevant:

- Continued spread from infected reindeer in Zone 2, Filefjell or any of the reindeer herds that have received animals translocated from Nordfjella to other animals in the same population and to neighbouring cervid populations. Enhanced surveillance, followed by adequate measures, will minimize this probability.
- Southward movement of CWD infected wild reindeer from Zone 2 to Hardangervidda and direct (unlikely) or indirect (more likely if mineral licks are present) spread of CWD to the main Hardangervidda population. If this occurred, it would escalate the challenges of CWD substantially. Enhanced surveillance and decreased population size, especially of reindeer males, will minimize this probability.
- Continued spread from infected red deer, moose or roe deer in the populations around Zone 1. The probability of presence of such cases in the large red deer populations in the western part of the area, having a very high density and documented space use and visits of mineral licks within Zone 1, both before and after the eradication, must be regarded to be relatively high in this context. If undetected spread of CWD within these populations occurs, further spread to new areas has to

be expected due to extensive seasonal migrations. Enhanced surveillance combined with population reduction will minimize this probability.

- Since absence of CWD infected cervids in the periphery of Zone 1 cannot be completely confirmed, establishment of new mineral licks inside Zone 1 and continued use of mineral licks in Zones 2 and 0, might facilitate spread of the prion agents if carriers shedding infectious prions are present or reappear, with mineral licks being hotspots for disease transmission. Reduction in the number of mineral licks will decrease this probability.
- The combination of high numbers of sheep grazing in Nordfjella with access to mineral lick sites, particularly those in Zone 1, but also elsewhere, increase the probability of passive transmission via sheep both within Zone 1 and out of the zone to other areas of Nordfjella. The probability that this should cause transmission to cervids is regarded to be very low for each case, but the cumulative probability might be low to moderate, depending on (among other things) the number of sheep that are put on home pastures accessible for cervids. is not negligible.
- High numbers of "guest grazing" sheep in Nordfjella that are subsequently transported to their home range, increase the probability of long-distance spread of CWD.
- Inadequate cleaning and/or disinfection of equipment, vehicles or garment used in Nordfjella increase probability of mechanical CWD spread. The probability of this causing spread to susceptible cervids is regarded as low but cannot be neglected.
- Non-ruminant animals (and humans) visiting Nordfjella and transporting contaminated material either in gastrointestinal tract or as dirt in feathers or fur and/or on feet cannot be excluded but is not regarded to represent a high probability of transmission to susceptible cervids.

6 Data gaps

Hazard identification and characterization

- The level and distribution of environmental CWD contamination in Nordfjella is unknown.
- The environmental persistence of the CWD strain in Nordfjella is unknown, especially the interaction between the prions and the soil and rocks in Nordfjella and the impact of the variable and moist weather there.
- The range of species susceptible to the Nordfjella CWD strain is unknown. Precautionary measures suggest that all cervids should be regarded susceptible.
- The transmission rate of the current strain in different Norwegian species of cervids
- The disease's course of infection with the strain in Nordfjella in different species of cervids and in eventual other species that prove to be susceptible.

- The ability of sheep and other species to act as nonclinical carriers that shed (small amounts) of prions without developing disease.
- The persistence and infectivity of prions in plants.
- The true prevalence or absence of CWD in cervid populations that have had contact with wild reindeer from NF-Z1 or their excreta.
- The number of reindeer (wild or semi-domesticated) that enter Nordfjella Zone 1 is unknown (assumed to be very low).
- The numbers of red deer and moose that enter and/or cross into Nordfjella Zone 1 is unknown.
- Whether mineral licks in Nordfjella Zone 1 (or other areas) attract cervids and thereby increase the number of animals entering from surrounding populations is unknown.
- The susceptibility of non-cervid species, such as livestock and humans to the Nordfjella CWD strain is unknown. Epidemiological and experimental evidence from North-America and studies of similar CWD strains suggest that livestock and humans share a very low susceptibility towards CWD prions in general. However, with the advent of new strains, this must be carefully investigated in each case.
- Whether sheep or other species can act as nonclinical carriers that might shed (small amounts) of prions without themselves developing CWD, is not known.

Exposure characterization

- How many carcasses of animals that have died from or with CWD have there been in Nordfjella
- The amount of CWD prions in different areas and sites of Nordfjella
- How many wild red deer and moose are exposed to CWD prions and how often does this occur
- The infectious dose of CWD prions in various susceptible species
- The consequence of exposure to repeated small doses of CWD prions on disease progression
- The spatiotemporal movement patterns of reindeer males, red deer, moose and roe deer in the area
- The destiny of the semi-domesticated reindeer at Geitrygghytta

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8 Appendix I

8.1 Cervid populations in Nordfjella Management Area

Before 1900, the populations and distributions of cervids in Norway were at a historical minimum, due to a combination of human persecution, large carnivores, harsh climate and land use for agriculture and livestock production. Wild reindeer populations were probably as small as a couple of thousand animals in the entire southern mountain range. In the northern half of Norway wild reindeer was completely replaced by semi- domestic reindeer, whereas several of the wild populations in the southern half already had experienced substantial interbreeding with semidomestic herds. Since the turn of the 18th century several of the semidomestic herds in Southern Norway went feral, - resulting in the present mixture of populations either having a wild, mixed or feral/semi domestic ancestry (ref). Reindeer population numbers started to increase after the Second World War, and populations in Hardangervidda and Snøhetta were at peak densities in the 1950s and 60s resulting in substantial over-grazing. The management authorities have since attempted to keep populations at certain target population sizes in accordance with local habitat qualities and grazing conditions (ref).

Reindeer are to a large extent living above the tree-line and populations are therefore distributed within a matrix of mountainous and forested habitat where human infrastructure and disturbance also act to hinder reindeer movements between populations. Management is to a large extent adapted to these distribution patterns and administrative management borders do to a large extent follow the biological borders of each population and not municipality borders (as is the case for red-deer, roe deer and moose). At present wild reindeer are living in more or less closed populations where management by harvest aims to keep populations at locally and predetermined target population sizes.

Red deer survived in five small pockets along the west coast (Haanes, Roed, Flagstad, & Rosef, 2010) (Langvatn 1999; Haanes et al. 2010). Moose survived in the inland closer to the Swedish border, while roe deer were extinct from Norway and only about 100 individuals survived in 1850 around a protected estate in south of Sweden (Cederlund & Liberg 1995). An expansion of cervids after this period was mainly driven by changes in harvest management, though extermination of large carnivores and changes in land use, both forestry (for moose) and agriculture (for red deer), play important roles. Roe deer and moose had a particularly rapid expansion in their distribution in Norway from 1950 and onwards (Andersen et al. 2004), but have now reached most of their potential range. An exception is for roe deer and moose along the central west coast. Today, mainly red deer is still expanding and at a slower colonization rate due to male-biased juvenile dispersal. Red deer populations are growing in numbers in the southeast, in the east and towards north of Norway. The distribution of roe deer towards mountains and further north are variable over time due to their sensitivity to deep snow and predation by lynx. They mainly colonize areas

near Nordfjella in periods with high population density in more suitable areas down in the valleys.

There is ongoing research to document both space use and basic population ecology of cervids in Nordfjella. Here, only a preliminary overview at a coarse scale is given. A more detailed overview is planned as part of a project ordered from the Norwegian Environment Agency.

8.2 Distribution and population development

Since the 1960s the wild reindeer management regime has aimed to keep population densities at predefined densities in accordance with current knowledge of pasture resources and population performance. Collection of data on population numbers was initiated during this period and has since the 1980's been gathered in annual censuses in Hardangervidda, and Nordfjella wild reindeer areas. Although no standard methods for estimations of population size has been implemented, annual surveys comprising minimum counts in winter, calf censuses in spring and demographic composition counts in fall, makes it possible provide rough estimates of population status and development trends.

In the other cervids a rough estimate of population densities and distribution at a coarse spatial scale, the municipality, were derived from the harvest statistics (from Statistics Norway). Over longer times scales harvest numbers give a reflection of population development of both red deer (A. Mysterud et al., 2007), moose (Ueno, Solberg, Iijima, Rolandsen, & Gangsei, 2014) and to some extent roe deer (Grotan et al., 2005; A. Mysterud & Ostbye, 2006). The exception will be during marked changes in harvest management. For example in Lærdal in 2017, the marked increase in harvest of red deer is not due to an increase in abundance, but reflecting increased harvesting to lower population density due to risk of CWD spillover. Harvest data give also a good overview of distribution. Harvest is typically initiated quite soon after colonization, though very high increases in harvest rates in early years suggests some delay between colonization and the onset of harvest (Milner et al., 2006). No data are available from year 2008. To get information about population densities, harvest numbers are divided by the 'qualifying area' used for setting harvest quotas, which typically is the area of suitable habitat within each municipality, which is mainly forest in the case of roe deer, moose and red deer.

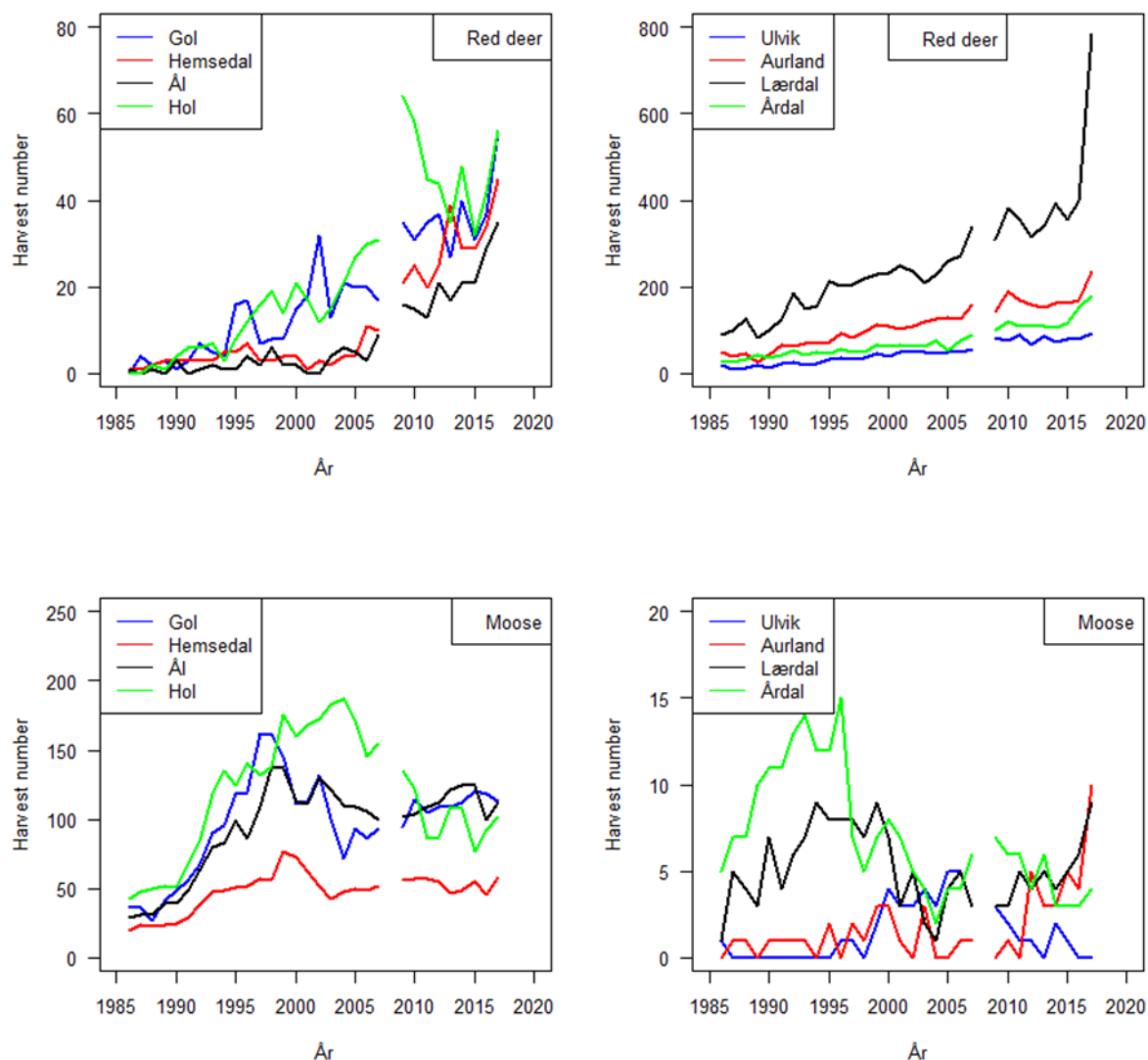


Figure 8-1. Harvest numbers of red deer and moose in the east (left column) and west of Nordfjella (right column). Note the much higher abundance of red deer in the west compared to in the east, and the opposite pattern for moose. Note different scale axes. Over longer time span, the harvest number will reflect population development, but not during periods with marked changes in management as for Lærdal in 2017 that will lead to population decline. Harvest numbers for roe deer are very low in the east and none are reported shot on the west side of Nordfjella during the period.

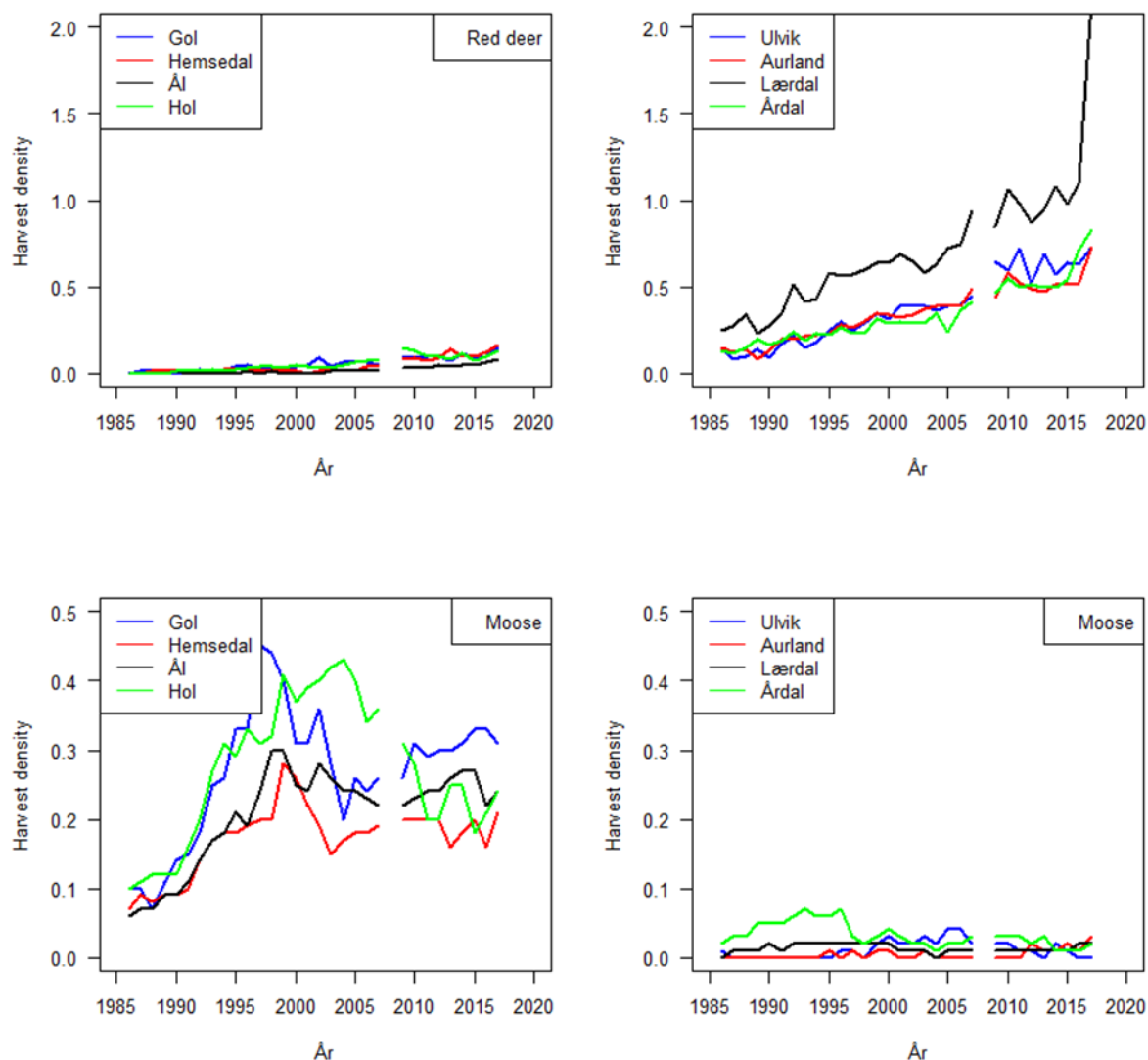


Figure 8-2. Harvest density (/km²) of red deer and moose east (left column) and west of Nordfjella (right column). Note the much higher densities of red deer in the west compared to in the east, and the opposite pattern for moose. Note different scale axes for moose and red deer. A rough rule-of-thumb to come from harvest density to (preharvest) population density is to multiply by 5 for red deer and by 4 for moose.

8.2.1 Movements and migrations

We classify movements of cervids and their drivers depending on the spatial scale. Broader scale movements relate to dispersal and seasonal migration. The drivers of dispersal and seasonal migration differ from each other and also from those of shorter daily foraging movements within seasonal home ranges.

Dispersal is mainly restricted to yearlings (or sub-adults) that leave their maternal home range without returning, typically in early summer. Dispersal is frequent for yearlings and often long-distance. Dispersal is more common among male red deer (Loe, Mysterud, Veiberg, & Langvatn, 2009), but occur at similar rates among sexes of roe deer (Wahlstrom & Liberg, 1995; Wahlström & Liberg, 1995) and moose (Debeffe, Rolandsen, Solberg, & Mysterud). Dispersal has limited directionality and can be difficult to predict (Loe et al., 2009). Dispersal was important for long-distance spread of CWD in USA (Lang & Blanchong, 2012), but represent a small proportion of the population and will not be discussed further in this report.

Seasonal migration is the main type of movements of cervids in and out of Nordfjella during an annual cycle, and this will be the focus. Migration is defined as non-overlapping space use between summer and winter. Migration is mainly elevational movements for all cervids, from low elevation winter ranges to higher elevation summer ranges (Bischof et al., 2012; A. Mysterud, 1999; Atle Mysterud et al., 2011; Rolandsen et al., 2017). There is hence a seasonal pulse of range contraction during winter (mainly due to snow) and a range expansion during summer as snow melt opens more suitable habitat (A. Mysterud, 2013). For red deer, migration also involves movements from winter ranges along the coast to summer ranges further inland (Albon & Langvatn, 1992; Bischof et al., 2012; Atle Mysterud et al., 2011).

8.2.1.1 Reindeer

Reindeer have adapted their behavior and physiology to a fluctuating and harsh environment. Most important is their ability to live in large herds. Both sexes carry antlers, and they form sexually segregated herds in late winter, spring and summer.

In contrast to the territorial behavior seen in other cervids, reindeer form temporal harems defended by competing males in larger herds during the rut. Males shed their antlers after the rutting season, whereas females keep their antlers and utilize the social rank and weapon they provide for food competition during winter. The most large-bodied females with large and symmetrical antlers have a competitive advantage. Males lose their social position after shedding their antlers and form relatively small buck groups that leave the main herds in early winter. Females, on the other hand, stay in larger groups also including younger animals and calves who follow their mothers until next calving season in May. During this period females normally depend on lichens as main winter forage. Since lichens are rich in carbohydrates but poor in protein and fat, females suffer from an energy deficiency in winter (starting approximately in February) and depend on access to the first green pastures in spring to provide sufficient nutrition for milk production peaking in the first weeks after calving. Female reindeer can lose as much as 20-30 percent of their body mass in harsh winters and can terminate pregnancies or alternatively lose their calves after birth if winter conditions are bad and calf birth weights too small.

8.2.1.2 Red deer

Red deer has been marked with GPS-collars in three different projects around Nordfjella. One project marked 17 adult female red deer in Buskerud during the period 2009-2010 (A. Mysterud, Bischof, Loe, Odden, & Linnell, 2012). In the project Hordahjort, 11 red deer were marked during 2009-2011 in Ulvik in Hordaland and Lærdal, Årdal and Aurland in Sogn & Fjordane. There is also an ongoing GPS-project called "Red deer in Nordfjella" with 30 red deer marked during winters 2016/17 and 2017/18 in Hol, Buskerud and in Lærdal, Aurland and Årdal in Sogn & Fjordane (Meisingset & Mysterud). The first study found that 94% of the 17 red deer were seasonally migratory in Buskerud (A. Mysterud et al., 2012). The proportion of migrants in Sogn & Fjordane is lower and in the range of around 70% (of 11 investigated animals?) overall (Atle Mysterud et al., 2011). Average migration distances are 30-40 km, but up to 100 km, and typically shorter closer to the coast in the west than in the inland. We recorded red deer marked in Lærdal with summer ranges far inland on the eastern side of Nordfjella (Meisingset & Mysterud). This likely follows the colonization routes from west to east.

8.2.1.3 Moose

The moose project in Hallingdalen marked 40 moose (29 female, 11 males) with GPS-collars during the period 2014-15 (Solberg, Rolandsen & Heim 2018). The proportion of migratory moose was similar among the sexes and around 60-70%. Spring migration was during April and first half of May, while the fall migration period lasted much longer with a mean at 9. October and a median date at 13. November. Migration distance was on average 25 km for males and 20 km for females.

8.2.1.4 Roe deer

A total of 19 roe deer was marked in Vestfold, Telemark and Buskerud, including some in Gol municipality on the eastern side of Nordfjella. In contrast to red deer, there were few migratory roe deer, and most roe deer remain at low elevation close to the agricultural areas year round (A. Mysterud et al., 2012). The lower proportion of migratory roe deer compared to red deer is a general pattern across Europe (Peters et al., 2018).

8.2.1 The role of salt licks for cervid distribution

There is little quantitative information on the role of natural and artificial salt licks for cervids space use from Scandinavian ecosystems. Soil ingestion at natural mineral licks (Kreulen 1985; Klaus & Schmid 1999), use of road salt (Tiwari & Rachlin 2018), antler consumption (Gambín et al. 2017) and even predation (Bazely 1989) by cervids to obtain minerals are well-known phenomena in general. That deer select minerals according to nutritional requirements is known from experimental studies of red deer in Spain (Ceacero et al. 2010). Studies of species like white-tailed deer (Atwood & Weeks 2002), sika deer (Ping et al. 2011), and moose (Tankersley & Gasaway 1983) show seasonal variation in use of salt licks

and patterns of use differ among males and females. Mineral composition of natural salt licks can benefit mineral intake (Ayotte et al. 2006), and they were frequently used during summer by wapiti and moose in Canada (Ayotte, Parker & Gillingham 2008). Licks are mainly used from spring to fall, and particularly in summer when body growth and reproductive costs are the highest. In Canada, moose may use salt pools during spring and summer in areas along highways (Laurian et al. 2008). Anecdotal observations in Norway suggest that cervids visit roads to lick salt during autumn and winter when road agencies spread salt on the roads to avoid icing (C.M. Rolandsen, pers. comm.).

Salts are usually not regarded as a limiting resource for cervid populations in Norway. However, given that artificial salt licks are available within their summer home range, we find it likely that this may affect the space use of animals. We find it less likely that salt licks affect the choice of summer home range and its boundaries. During summer, boundaries of cervid home ranges are partly determined by social factors and kinship. Movements outside of ordinary home ranges are not common. We therefore regard it unlikely that salt licks for sheep in Nordfjella attract an additional number of cervids to the region. However, this is based on expert opinion and further quantitative studies of cervids use of salt licks are needed for confirmation. Red deer and moose will likely mainly use salt licks positioned in the forested areas. During nighttime, both red deer and moose use alpine areas above the treeline to some extent and likely may use salt licks a few km into open alpine habitat.

8.3 Movements of semi-domestic reindeer into Nordfjella Nf-Z1

There are no GPS-marking data available on semi-domestic reindeer in Filefjell, which is the adjacent population to Nf-Z1. However, there is annual reporting of the number of semi-domestic reindeer shot within Nf-Z1 (Table 4-1). The annual average was 4 semi-domestic reindeer shot (not including 2017), which likely represents an underestimate as not all hunters are aware they should report this.

The influx of semi-domestic reindeer to Nf-Z1 is regarded a critical risk for a new CWD epidemic. The fences along the road over Hemsedalsfjellet is unlikely to yield an effective year-round barrier due to snow deposition (Mysterud & Rolandsen 2018), but as explained above the semi-domestic reindeer herd do predominantly use the areas along the road for summer pastures. It is uncertain how many animals enter and then exit Nf-Z1, which would be the critical parameter from a disease containment perspective, but according to Filefjell reinlag the current fences and their herding routines should result in a very low number of such episodes.

Table 8-1. An overview of the number of semi-domestic reindeer shot in Nordfjella zone 1 during 2008-17. Data retrieved from Aurland Fjellstyre (provided by H. Skjerdal)

Year	2008	2009	2010	2011	2012	2013	2014	2015	2016	2017
Number felt	5	2	8	7	0	0	6	5	4	30

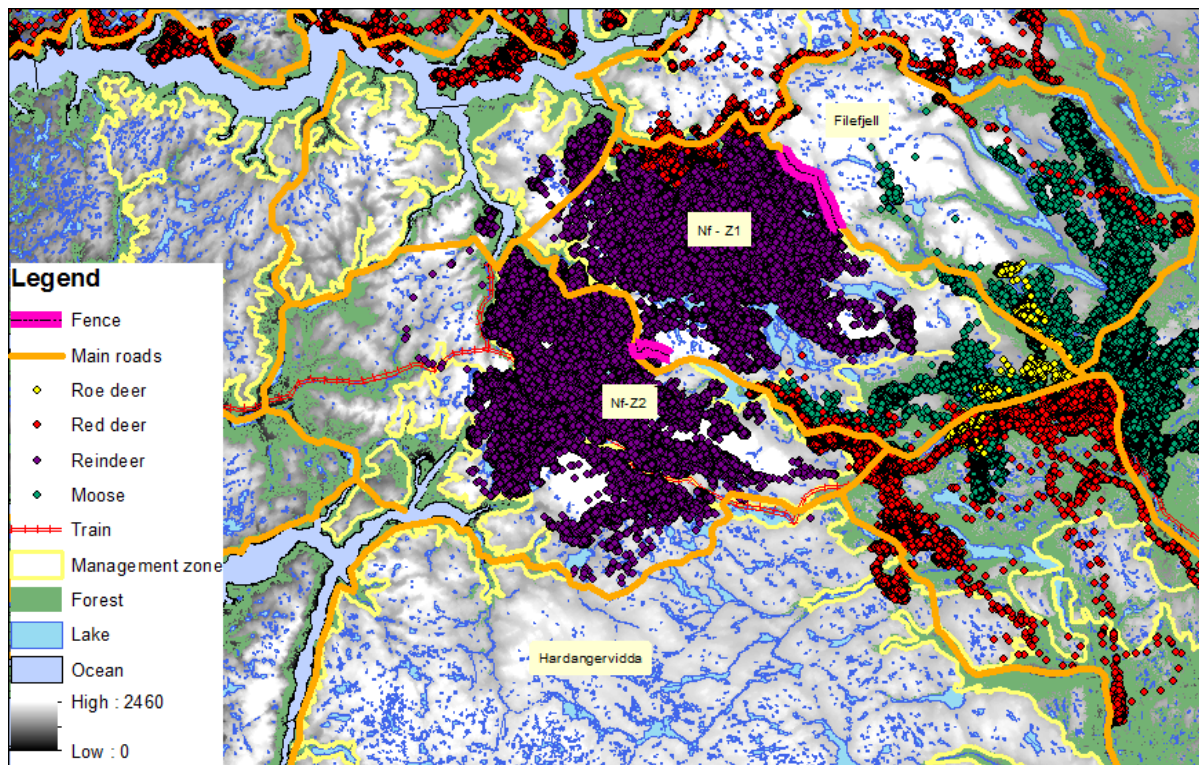


Figure 8-3. Examples of spatial overlap of wild cervids in the Nordfjella region as evidenced from GPS-data from reindeer in NF-WRA, red deer, moose and roe deer. There is substantial overlap for example between red deer and reindeer at the northern margin of NF-Z1. The pink lines illustrate the new reindeer fences build on crossing points along FV50 and RV52. Note that while the reindeer positions give a relatively good picture of the space use of a whole herd of female reindeer, the distribution of the other positions represent different individuals' space use. Consequently, absence of positions does not necessarily mean that no animals of a species utilize that habitat, but can just as well mean that no animals were marked in that area. Be aware that the space use of wild reindeer in the Hardangervidda population, wild reindeer in the Lærdals-Årdalsfjella population and semidomesticated reindeer in Filefjell not are shown in this figure. (Dataowners; O. Strand (reindeer), E. Meisingset & A. Mysterud (red deer), E.J. Solberg & C.M. Rolandsen (moose) and A. Mysterud & J.D.C. Linnell (roe deer).)

9 Appendix II

9.1 Sheep farming in Norway

Only about 3% of Norway is cultivated agricultural land, whereas massive areas are well suited for summer grazing. This eco-friendly resource, combined with fisheries, has been of pivotal importance for human settlement and survival in Norway for centuries. Even today, these mountain resources are important for the Norwegian agricultural industry and it is a deeply interwoven element of rural culture.

The use of mountain pastures for sheep grazing has long traditions in Norway and sheep farming is a common production in many remote and rural areas of the country. On a national level, the number of sheep has increased the last decade and the total population was 2.417.813 in the 1st of October 2017 (Statistics Norway). The number of farms with sheep production, however, has decreased in the same period, and the average size of the herds consequently increased. In general, ewes are kept in barns during the late autumn and winter. Several farmers keep a ram together and circulate it between the herds to fertilize their ewes, often organized into a system where the best rams are used (called “værringer”). The ewes give birth to lambs in the early spring (relative to greening). When the grass has sprouted, and the fields are green, the ewes and their lambs are let out on home pasture close to the farm, but as soon as the snow has melted, and grass sprouted also in the mountains, they are transported up to higher altitudes. Here they disperse out on the land, most often grazing in small groups. The sheep are not herded in a traditional way with herdsmen continuously watching them, but regulations require that there is some kind of supervision of the animals, at least once a week. Man-made mineral licks are often put out strategically to encourage the sheep to stay in the areas the farmer wants. All sheep are marked and many of them carry bells, to ease finding and herding. Many sheep now also carry radiocollars. In autumn, as the risk of getting snow is increasing, the farmers get together and gather the sheep down to corrals in a process called “sanking”, where they are sorted in herds and according to their destiny. The sheep can now be sent directly to the slaughter house, but it is also common to bring them home and let them graze for a while on the pastures around the farm before most of the lambs are sent to slaughter and some lambs and the sheep intended as a breeding stock for next year's production and housed in the stables again. Most of the users of the pastures are organized in an arrangement called “organisert beitebruk”. This was established around 1970 as a cooperation between the Ministry of Agriculture and the Norwegian sheep and goat farmers association, Norsk Sau og Geit. The aim was rational use of mountain and forest pastures and reduction of losses of sheep and lambs (Drabløs, D., 1997, Soga om smalen., Norsk sau- og geitalslag, Oslo, Norway and www.nsg.no accessed 1st of November 2018). This occur through organized supervision at pasture (Norw: “tilsyn”), cooperation during collection and gathering) in autumn and cooperative measurements as fencing, building of corrals, maintenance of sheds for herders and provision of mineral licks. Organisert beitebruk is facilitated by the

authorities, who for example provide grants for the measurements mentioned above, meant to decrease losses or ease the use of the pastures. Locally, the sheep owners are organized in "beitelag" or "sankelag" who organize the cooperation in a given area.