Oh Deer! Prion Diseases in Minnesota

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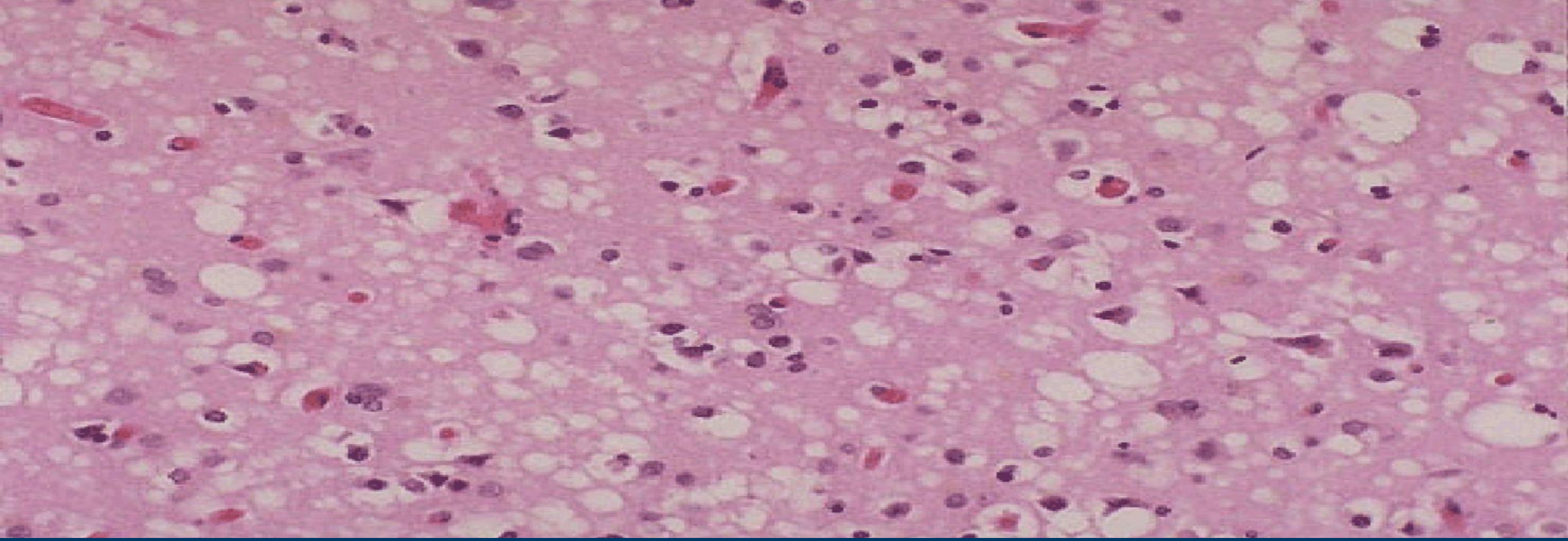
Responsibilities of ZDU

Agritourism and petting zoos
Animal bites
Animal contact (agriculture and household pets)
Anthrax
Avian influenza
Bartonella spp.
Blastomycosis
Botulism
Brucellosis
Campylobacter (acquired from animal contact)
Capnocytophaga canimorsus
Chronic Wasting Disease
Clostridium difficile
Coccidioidomycosis
Cryptococcus neoformans
Cryptosporidium (acquired from animal contact)
Cysticercosis
Shiga toxin-producing *E. coli* (acquired from animal contact)
Hantavirus
Histoplasmosis
Laboratory exposures
Leptospirosis
Melioidosis and glanders
New World Screwworm
Novel influenza
Orf and other parapox viruses
Orthopox virus
Pastuerella spp.
Plague
Prion Disease
Psittacosis
Q-Fever
Rabies (human and animal)
Rat Bite Fever
Raw milk
Salmonella (acquired from animal contact)
Swine influenza
Toxoplasmosis
Trichinosis
Tularemia
UMASH Center projects
Unexplained deaths and critical illnesses
Veterinary infection control
Viral hemorrhagic fevers
Zoonotic occupational exposures

Objectives

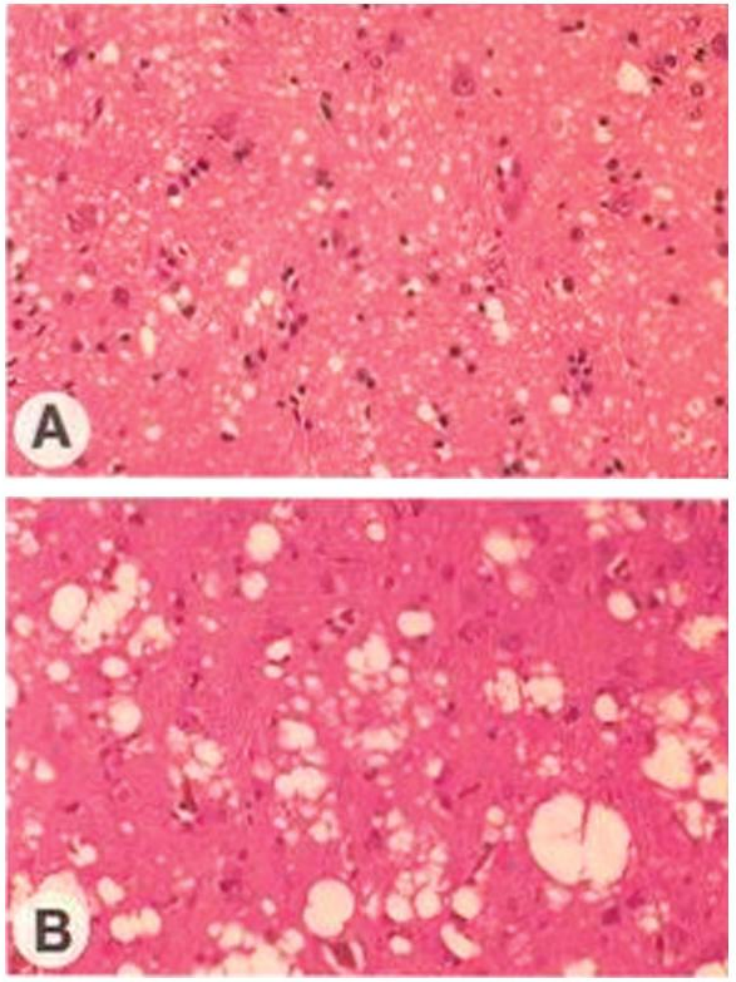
- Describe the major forms, epidemiology, and transmission routes of CJD and other prion diseases
- Assess the risk of zoonotic transmission of prion diseases
- Highlight the surveillance and epidemiologic investigations of prion diseases in Minnesota



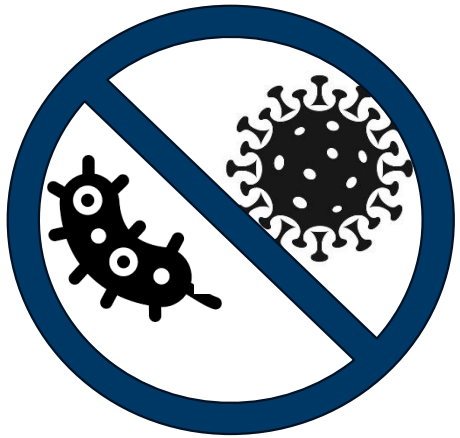
Minnesota Surveillance of Prion Disease

Transmissible spongiform encephalopathy (TSE)

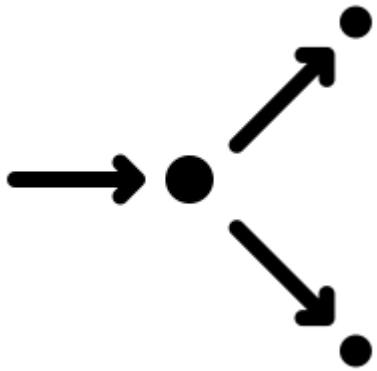
- Family of fatal diseases called TSEs
  - "Transmissible" – can spread from one individual to another
  - "Spongiform" – brain riddled with holes
  - "Encephalopathy" – disease of the brain
- Incubation period of months to years



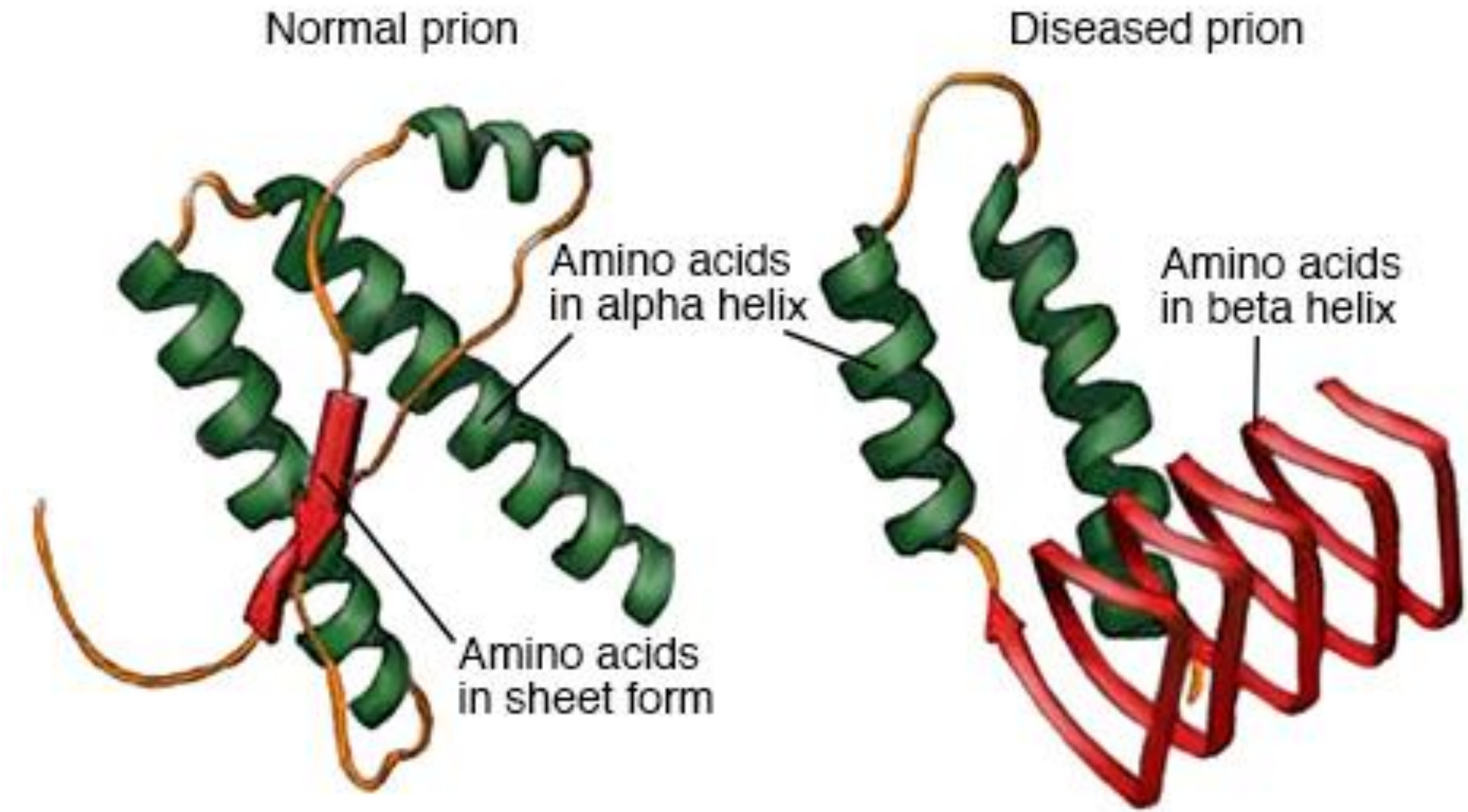
These diseases are caused by prions



- Prions are **protein**, that cause similar proteins in the brain to fold abnormally



- In a chain reaction, the diseased prion causes a normal brain prion protein to change shape and become a diseased prion



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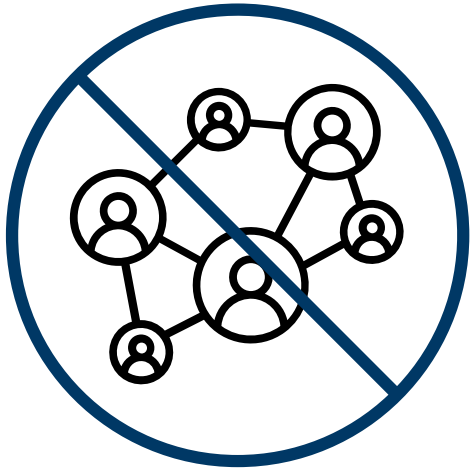
Relevant TSE biology

- Prions are nearly indestructible
  - Cooking does not affect their infectivity
  - Autoclaving, disinfectants, and none of the usual infection control processes are effective
- Heat to 1832°F, alkaline (NaOH or KOH) tissue digesting, and 50% bleach solution (on stainless steel) are effective

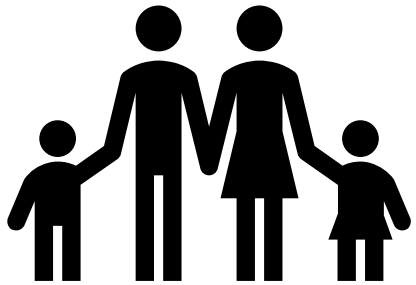
Sporadic TSEs in humans

- Estimated 85% of CJD cases
- Classic Creutzfeldt-Jakob Disease (CJD)
  - Six different subtypes
  - Different subtypes have different clinical presentation
- Sporadic fatal insomnia



Inherited TSEs in humans

- Less common, estimated 5-15% of cases
- Gerstmann-sträussler-scheinker disease (GSS)
- Fatal familial insomnia
- Genetic CJD

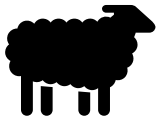


Acquired TSEs in humans

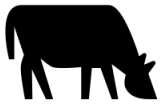
- Iatrogenic: caused by contact with prions via medical procedures or medical equipment
- New-variant CJD (vCJD): BSE in humans from eating BSE-infected beef
- Kuru: previously found in New Guinea from eating human brains as part of a funeral ritual



TSEs in animals



- Scrapie (sheep and goats)



- Bovine spongiform encephalopathy (Mad Cow Disease)



- Transmissible mink encephalopathy



- Feline spongiform encephalopathy



- Chronic wasting disease (CWD) of deer, elk and other cervids

BSE is the only TSE known to be zoonotic

- First identified in cattle in the UK in 1986
- First human case of vCJD identified in 1996
- 750,000 infected cattle entered human food chain before BSE control measures were instituted
- Over 220 human deaths occurred

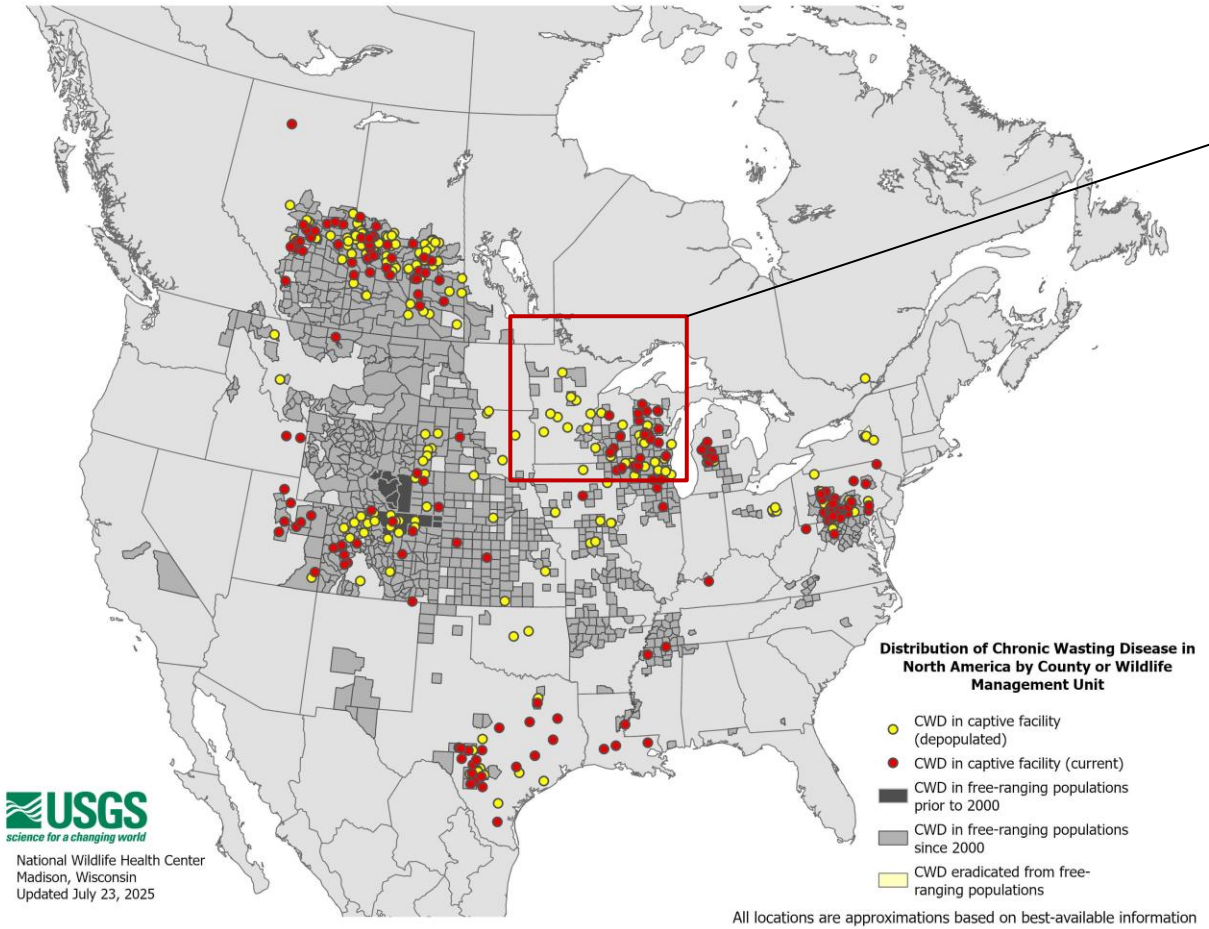


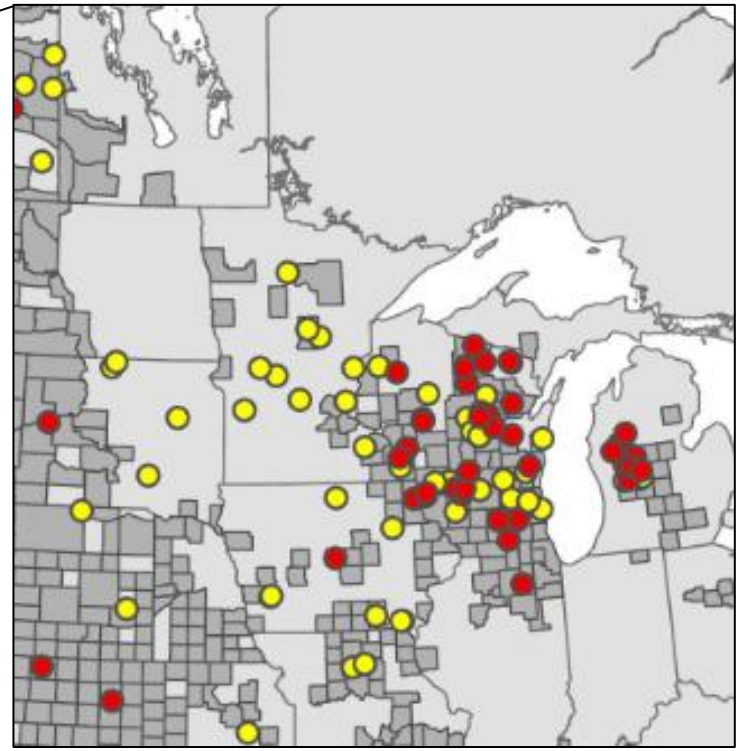
CWD is the only known TSE of free-ranging wildlife

- First identified in captive mule deer in a Colorado research facility in 1967
- Most infectious and easily transmitted of the TSEs
  - Found in saliva, urine, feces, semen
- Transmitted between deer through contact or the environment

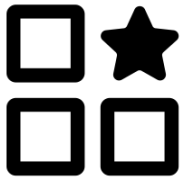


Distribution of CWD

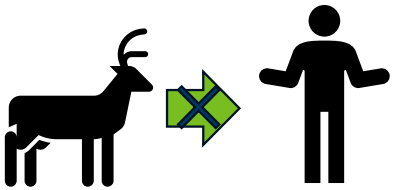




What do we know?



- CWD is unique. The known prion diseases of humans and domestic animals are different from CWD



- CWD has been known and studied for decades and there has been no evidence that CWD has been transmitted to humans



- CWD has not been identified in domestic animals (including dogs and cats) or livestock other than deer/elk

What do we know?



- In the laboratory, species barriers can be breached for all the prion diseases. A prion disease normally found in one species can be made to infect another species



- Some prion diseases that are adapted to a particular host species can naturally infect other species



- This is why MDH takes a cautious stance on the question of human health risk from CWD

Demographics

- Age at illness onset and death is an important factor
- 80% of cases of classical CJD are 50-70 years of age

vCJD from infected beef



Average age of onset:
29 years old

Sporadic CJD



Average age of onset:
60 years old

Disease course

- Because CJD is sporadic, there is not a known incubation time
- Patients may present with changes in mental ability
- CJD has a much more rapid progression than one may see with other brain disorder







Disease course (cont.)

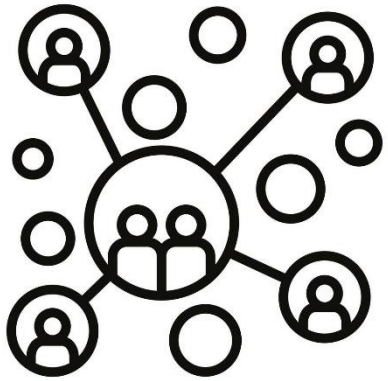
- Early symptoms
  - Memory loss, behavior changes, coordination issues, sleep problems, vision changes
- Advanced symptoms
  - Involuntary movements, severe cognitive decline, communication issues
- Death usually occurs within a year of symptom onset

What is MDH doing?

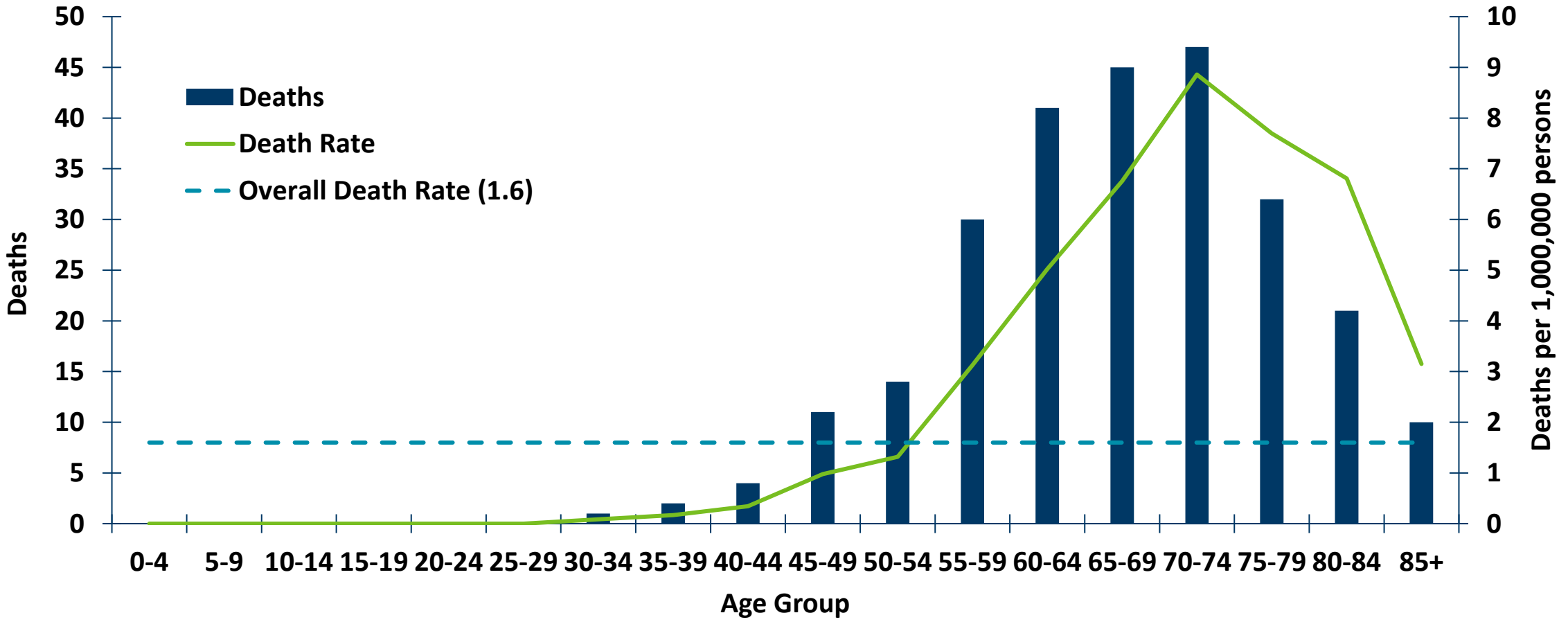


- Routine human prion disease surveillance – mainly CJD – to identify illnesses possibly related to prion diseases in animals

- We look for anything unusual
  - Clusters of disease
  - Atypical case-presentations
  - Unusual exposure histories
  - Unexpected age groups



Creutzfeldt-Jakob Disease Deaths and Death Rate by Age Group, Minnesota,1996-2022*



* Based on death certificate data, reports from National Prion Disease Pathology Surveillance Center and other healthcare providers; Minnesota population based on estimates made by the National Center for Health Statistics (NCHS)

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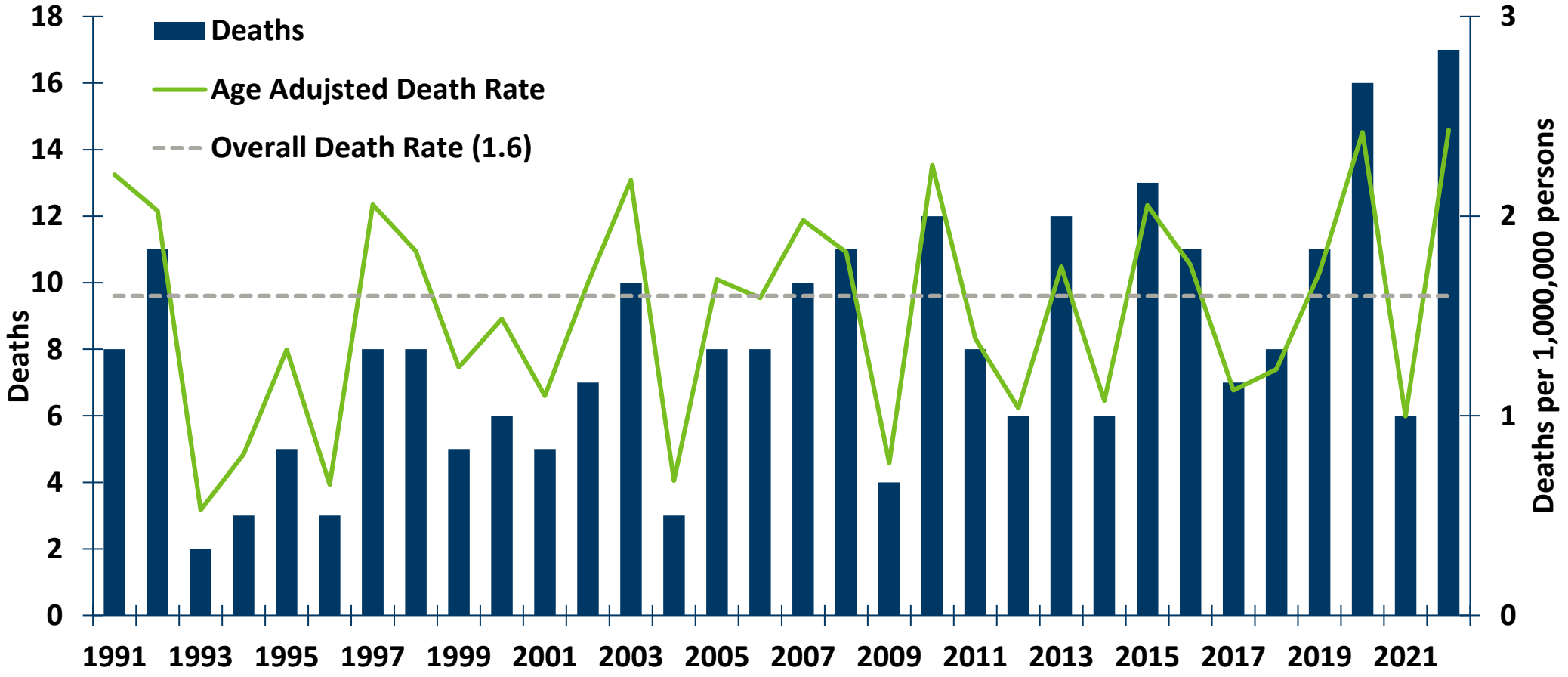
Creutzfeldt-Jakob Disease Deaths and Age-Adjusted Death Rate, Minnesota, 1991-2022*



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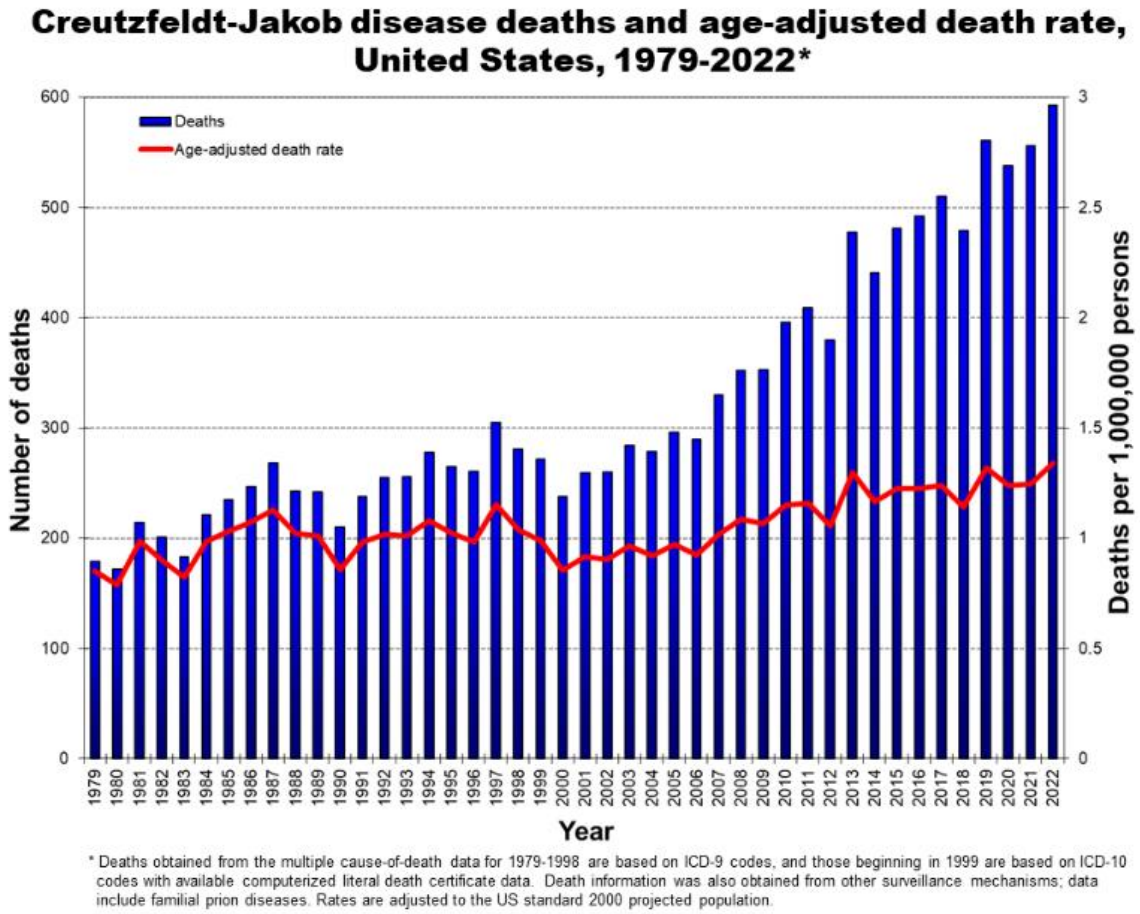
DEPARTMENT OF HEALTH

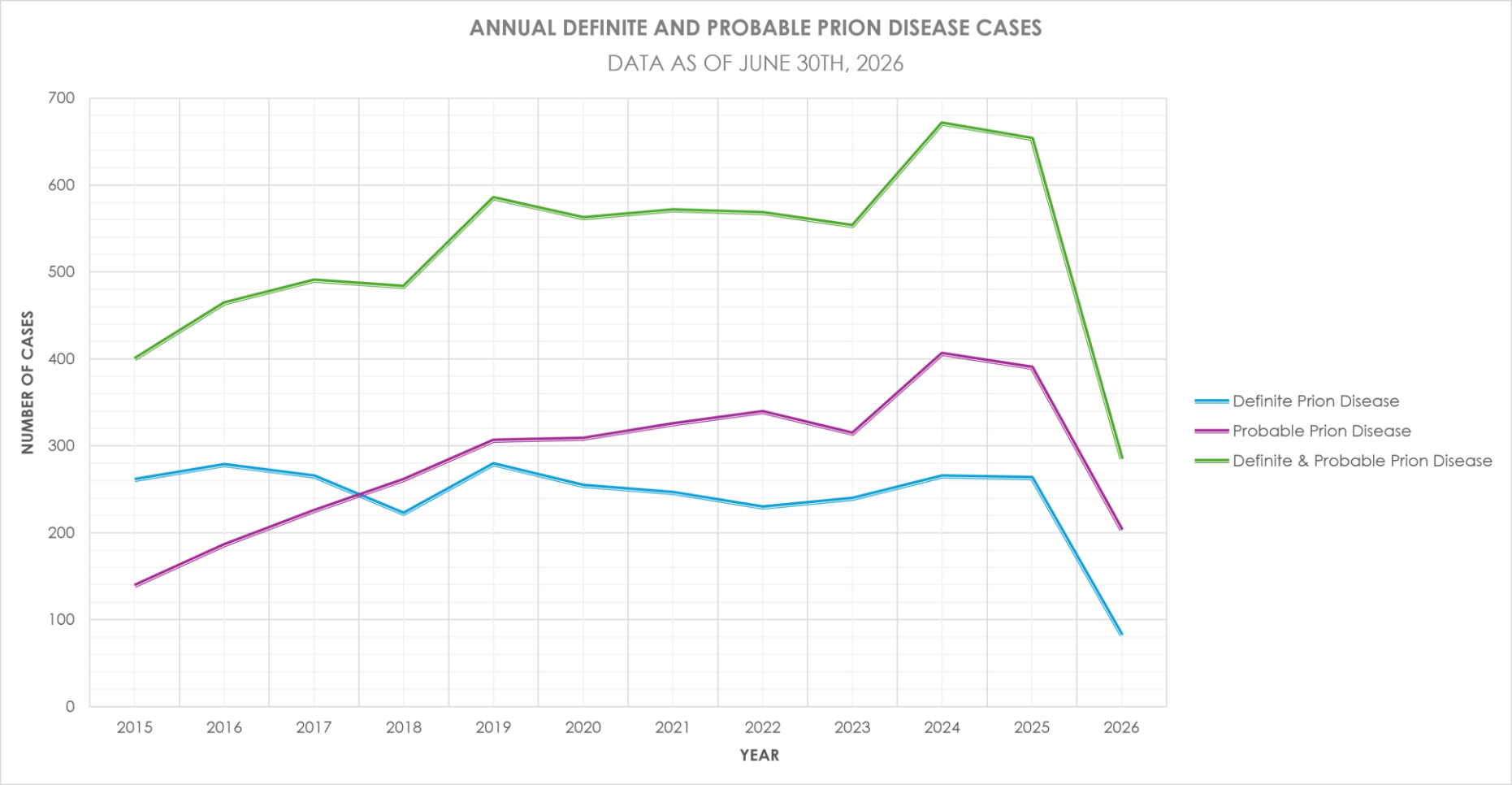
Prion disease in the US

- 500-600 cases of CJD are reported each year in the US
- No state has higher incidence of prion disease than others
- Some states report higher number of CWD positive cervids



Annual CJD deaths and age-adjusted death rate from 1979-2022.

National prion disease surveillance data on annual definite and probable prion disease cases



Conclusions:

- MDH has not identified any potential cases or case clusters that might indicate a new prion disease in humans
- Iatrogenic transmission from medical procedures or medical equipment is rare; but it is important to remain diligent about the transmission potential
- Prion disease can only be confirmed via autopsy – offered for free
- No evidence of natural CWD transmission to humans
    - Never-the-less, MDH does not want people eating prions from any infected animal of any species

Resources

- National Prion Disease Pathology Surveillance Center
- Mayo offers RT-QuIC testing
- CJD Foundation for families of those
- World Health Organization for Infection Prevention and Control
- MDH Zoonotic Diseases Unit epidemiologists

Thank You!

Contact Info:

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