

Alberta Prion Research Institute

Prion Diseases and Dementias

Kevin Keough PhD FCAHS ICD.D

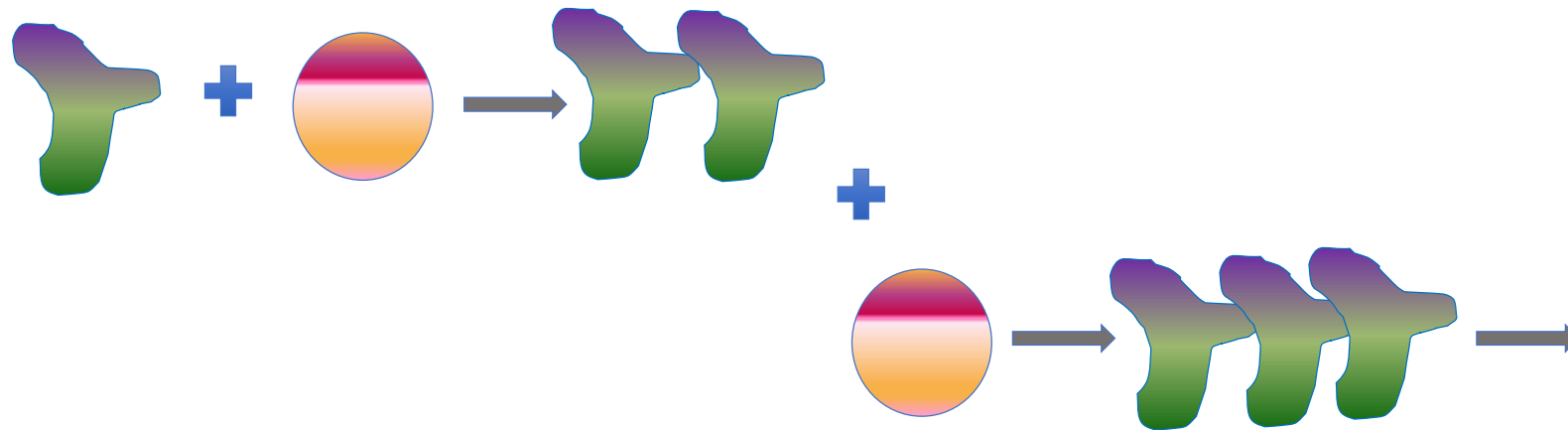
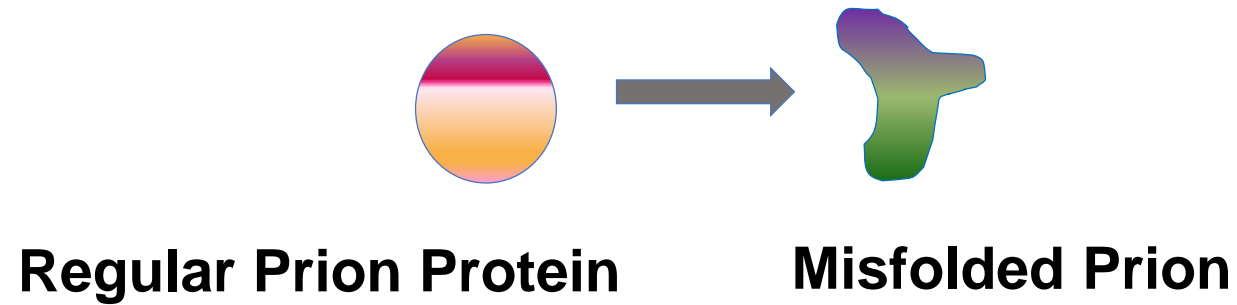
Executive Director

July 15, 2019

Why would you want to know about Prions?

- Animal Diseases
- Human neurodegenerative diseases and dementias

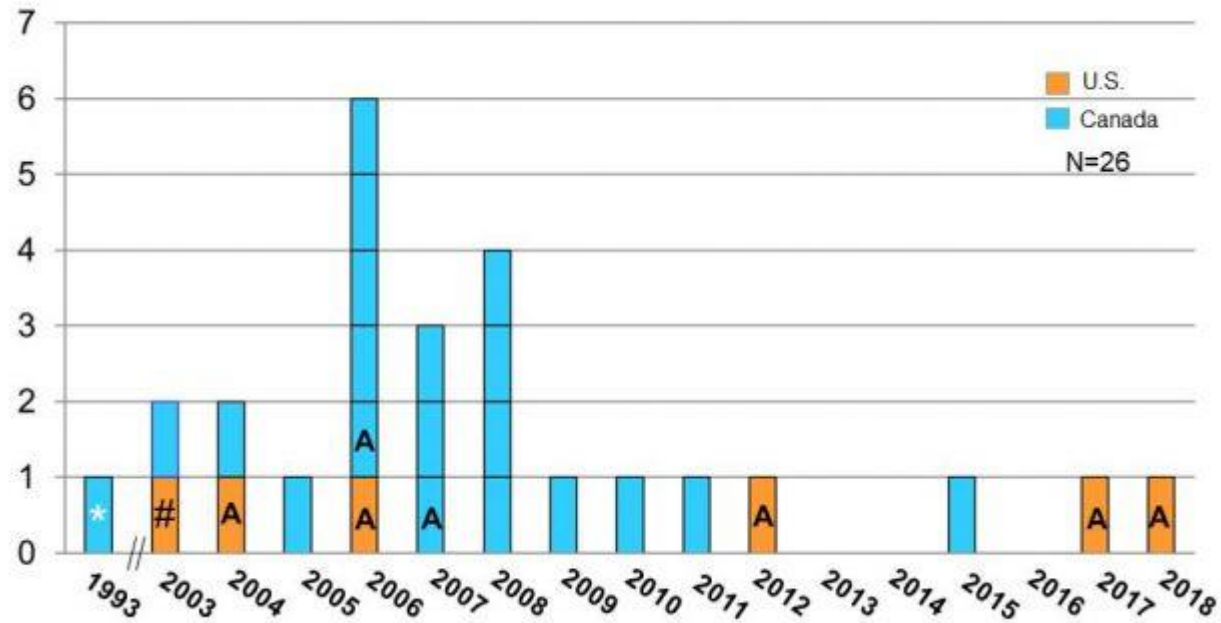
What is a prion?



Prion diseases

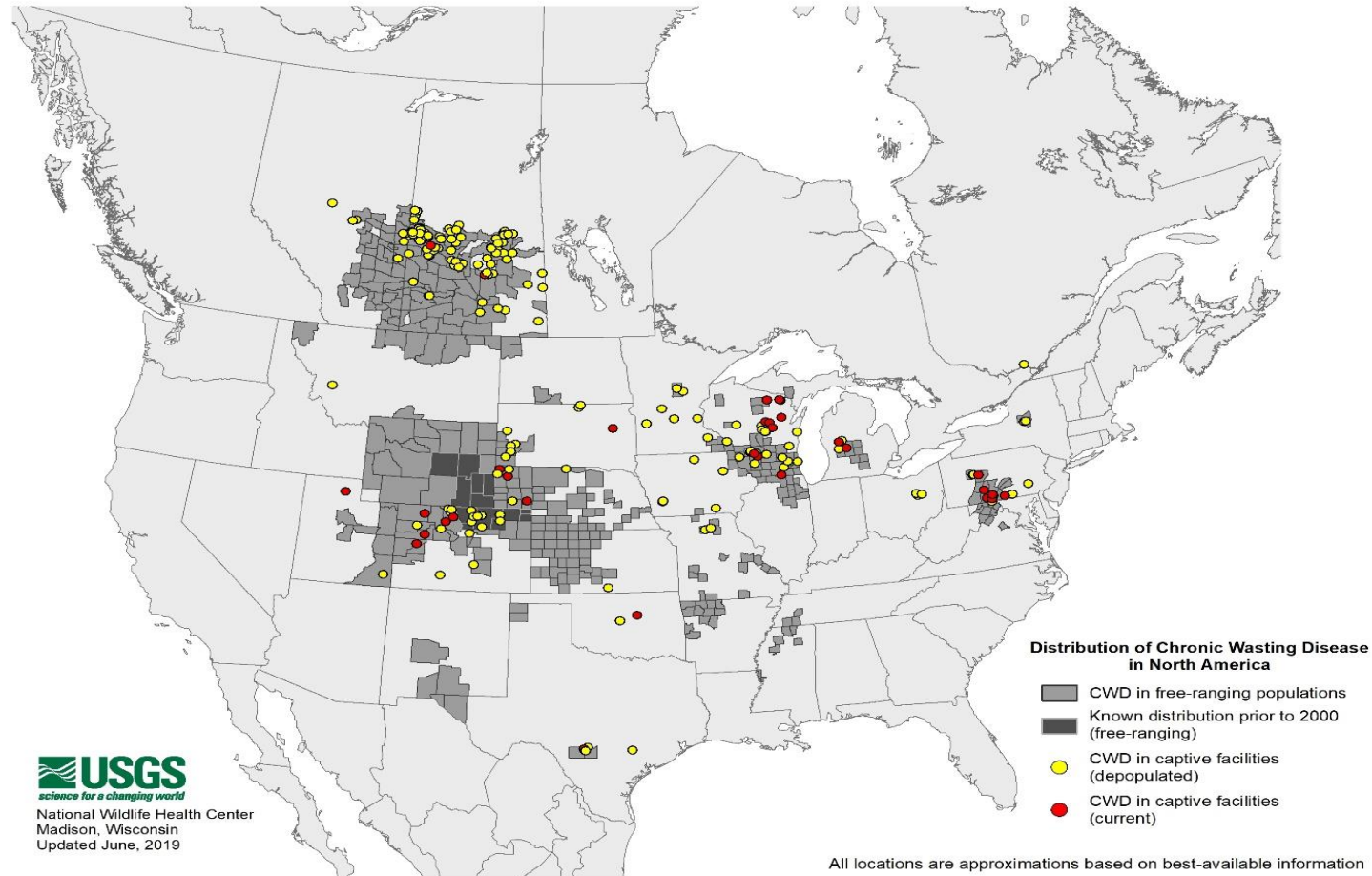
- Bovine Spongiform Encephalopathy (BSE)
- Chronic Wasting Disease (CWD)
- Scrapie
- Creutzfeldt-Jacob Disease (CJD)

BSE in North America

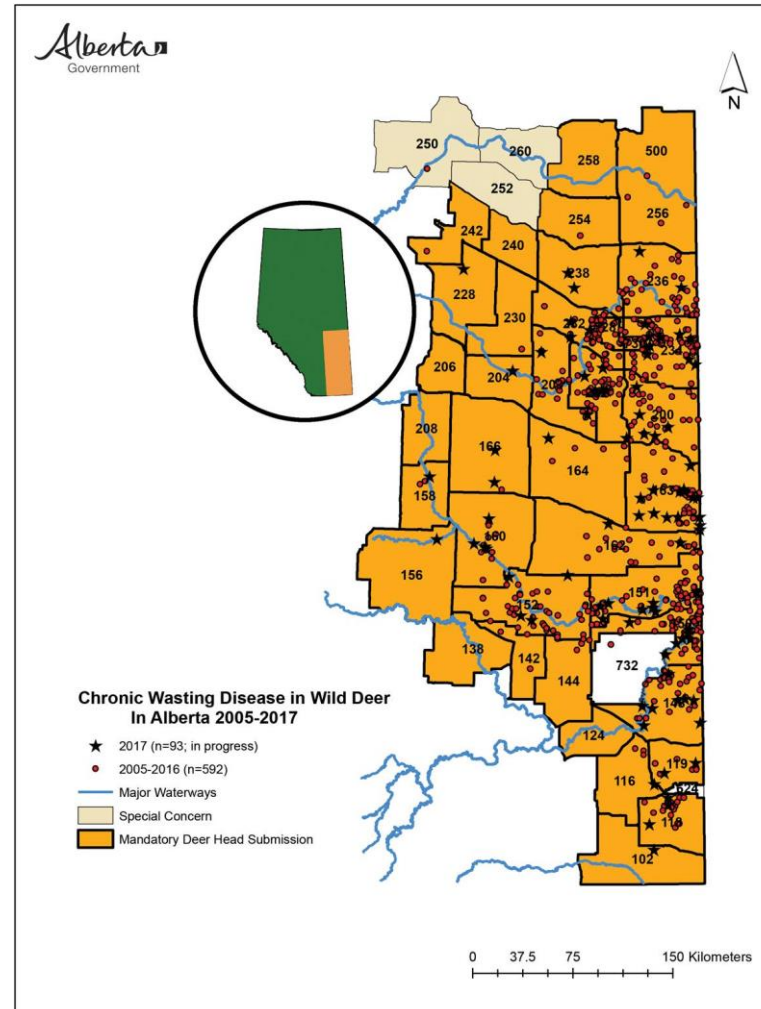


Key: A-Atypical strain of BSE, #-Imported, Canada to US, *-Imported, UK to Canada

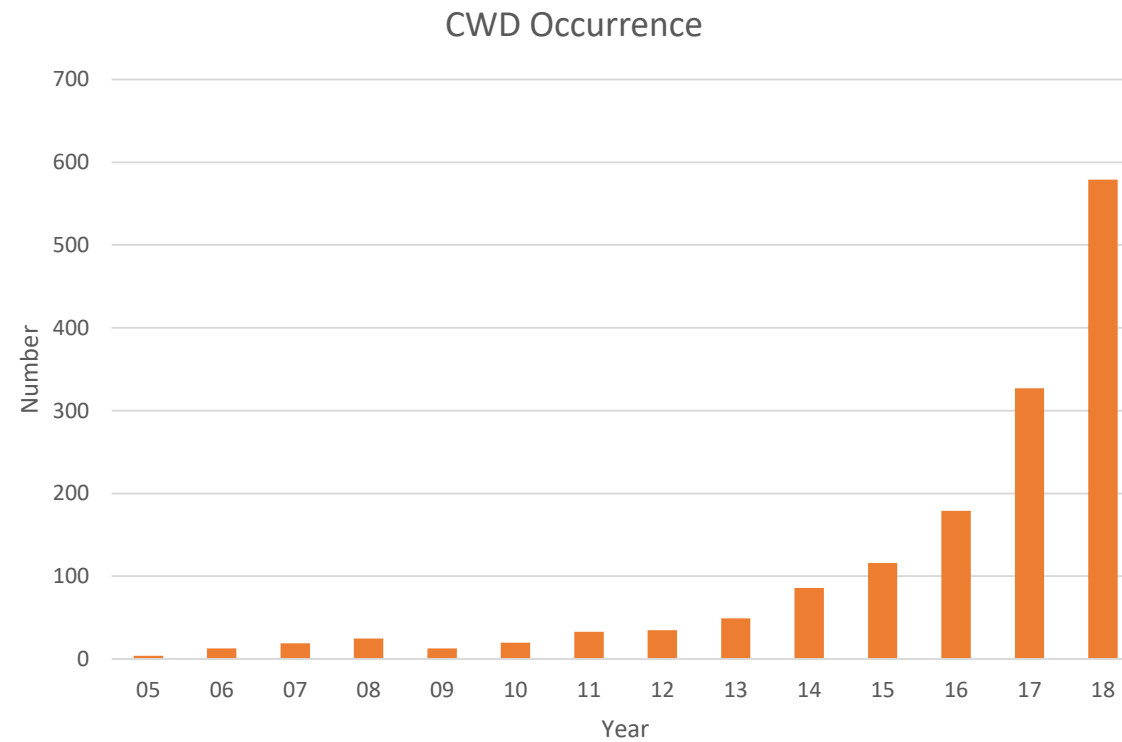
CWD in wild deer in North America



CWD in Wild Deer in Alberta



CWD Occurrence - Alberta



CWD transfer

•	Possibility	Probability
• Cervids	Possible	Very high
• Humans	Possible	Low
• Cattle	Possible	Low
• Pigs	Possible	Low - medium
• Sheep	Possible	Low

The crop challenge

- CWD prions can stick to and be taken up in plants
- CWD prions can stick to some soils
- These become sources of infections

- Norwegian action banning import of hay and straw from CWD endemic areas

Alberta Prion Research Institute

- Created in 2005 in response to the 2003 BSE crisis
- Funding research on animal prion diseases and human prion and prion-like diseases
- \$77.5 million over 16 years (to expire in March 2021)
- Over 450 young researchers and technicians trained in funded labs

Alberta Prion Research Institute



ALBERTA PRION RESEARCH CONNECTIONS

- NATIONAL
- - - INTERNATIONAL



Current BSE research

- Reducing the cost of removal of potentially BSE-contaminated materials
- Can new rare forms of BSE cause “classical” BSE
- Structure of BSE prions

CWD current research

- Transmission to non-human primates
- Transmission between cervids – at least 5 forms (strains) of CWD
- Management on landscape
- Diagnostics – finding CWD in animal droppings (patent)
- Vaccines – rational vaccine design (patent)
- Therapeutics

Prion-like diseases

- Alzheimer's disease
- Parkinson's disease
- Frontotemporal dementias
- Tauopathies
- Amyotrophic lateral sclerosis
- Other dementias

Dementias and Alzheimer's Disease

- Approximately 750,000 people with dementia in Canada
- Approximately 82,000 people with dementia in Alberta
- About two-thirds of dementias are Alzheimer's Disease

Current Alzheimer's disease research

- Searching for therapeutics
 - Artificial nerves for testing drugs (patent)
- Biomarkers and diagnostics
 - Blood test for early detection of Alzheimer's disease (patent and company)
- Fundamentals of what is going wrong
 - Different forms (strains) of Alzheimer's disease agents

A few of our funded researchers



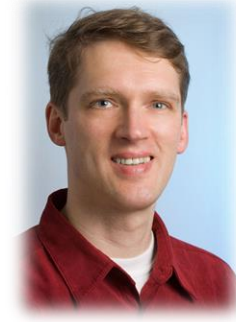
David Westaway



Stefanie Czub



Peter Stys



Holger Wille



Sabine Gilch



Ted Allison



Debbie McKenzie



Valerie Sim

Partners and supporters

- Provincial, national and international government departments and agencies
- Universities
- Industry
- International research agencies
- Alzheimer Society of Alberta and Northwest Territories

Alberta on the international scene

- Largest group of prion researchers in the world
- Recognized as the leading jurisdiction
- Opportunity to build on prion investment to be a research leader in diseases such as Alzheimer's

- Thank you