# VMC 321: Systematic Veterinary Virology

# Prions

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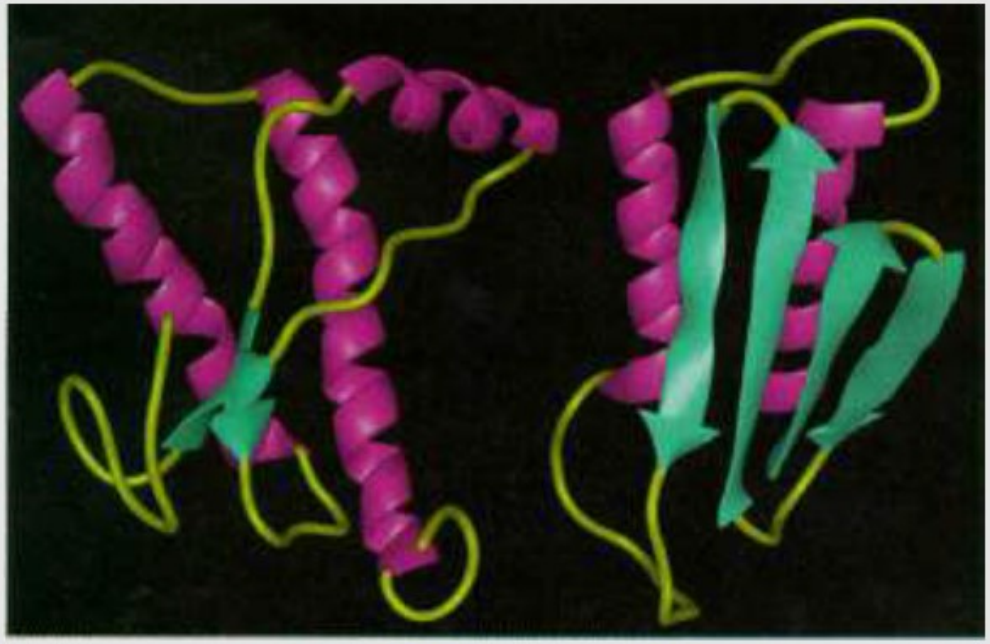
# Prions

- Prion - Pronounced “pree-on”
- Shortened term for: Proteinaceous Infections Particle
- The word “prion”, was discovered and coined by Prusiner in scrapie.
- Prion means a proteinaceous infectious particle
- Prions ($PrP^{Sc}$) are infectious proteins
- Unique in their ability to reproduce on their own and become infectious.
- Cause neurodegenerative diseases in humans and animals.

# Prions

- are acellular entity
- proteins found mostly abundant in the brain
- doesn’t use energy
- doesn’t grow
- prions don’t die
- they reproduce via pre-existing prions

# Prion - Basic structure

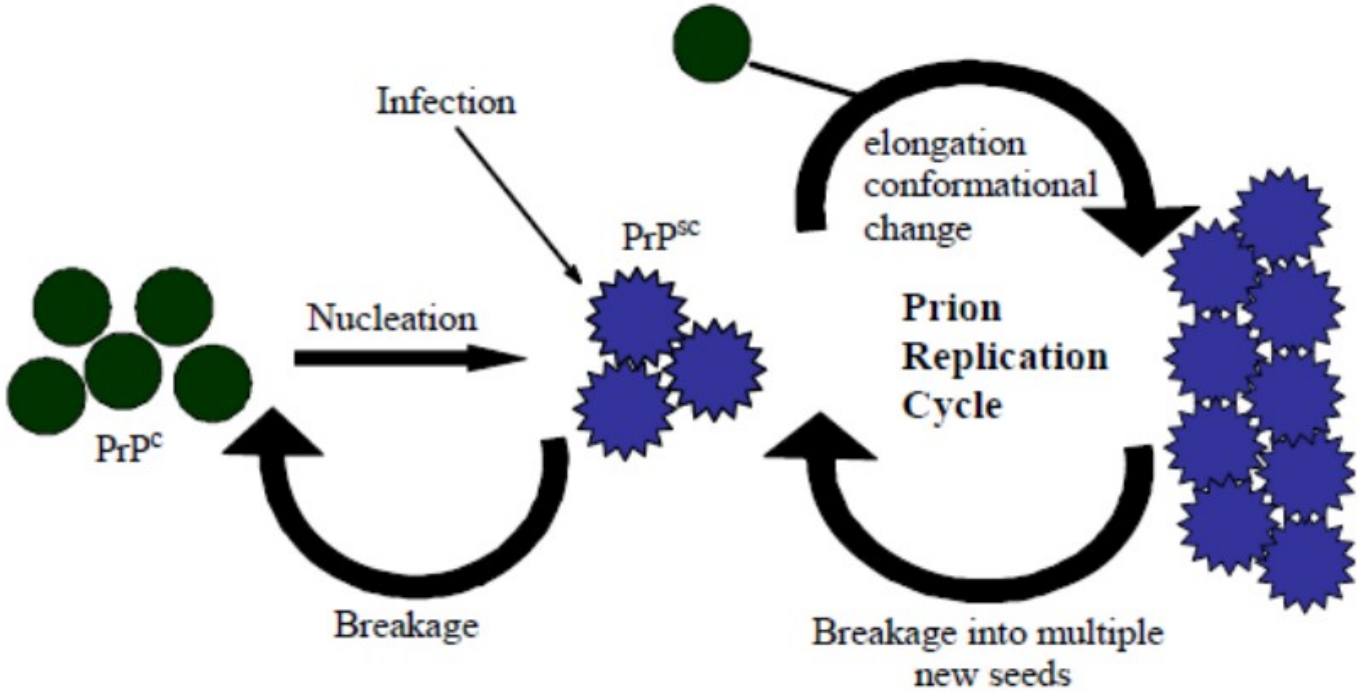


Normal Mutated

- Normal prions
- contain about 200-250 amino acids
- twisted into three telephone chord-like coils known as helices, with tails of more amino acids.
- The mutated, and infectious, form - built from the same amino acids but take a different shape.
- 100 times smaller than the smallest known virus.

# Replication

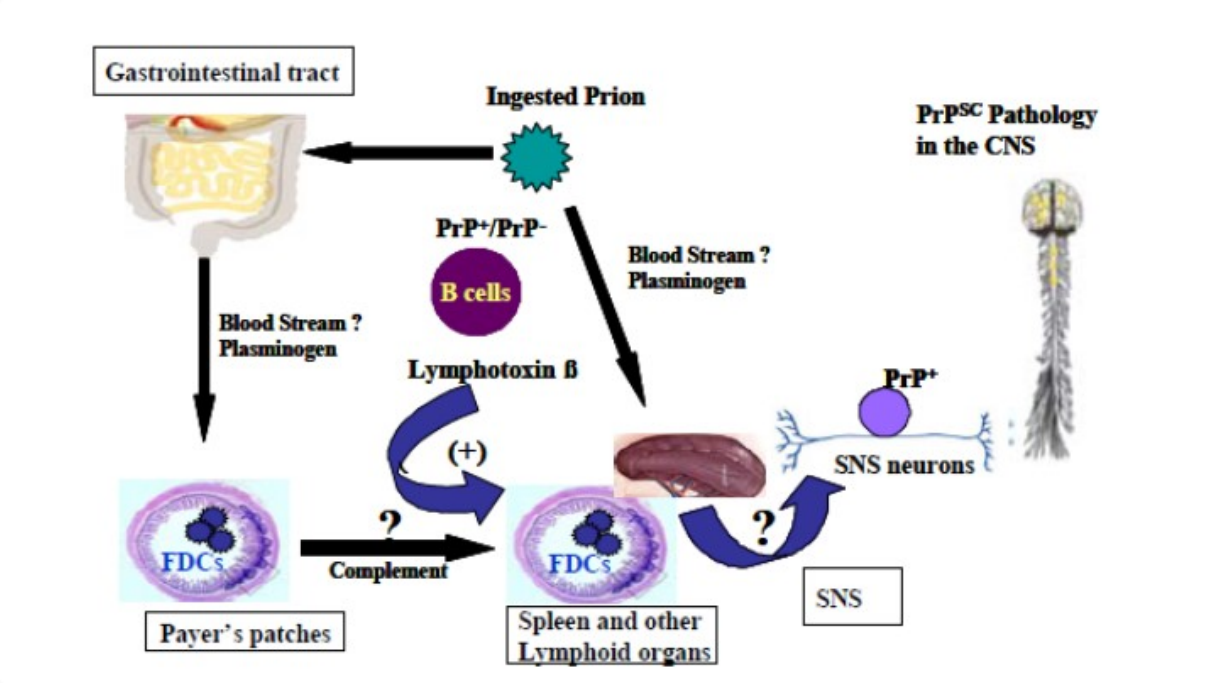
## Prion Replication Cycle



# Pathogenesis

- prions silently accumulate and replicate in peripheral organs or reservoirs
- transit through at least one PrP-positive (PrP+) tissue before reaching the CNS.
- Prions replicate in lymphoid organs during the early stages of infection
- Within the lymphoreticular system, follicular dendritic cells (FDCs) are a prominent site of $PrP^{sc}$ deposition

# Route of Prion Neuroinvasion




- absorption through the intestinal epithelium
- prion reach the peyer's patches, via blood constituents (Plasminogen that bind PrPsc).
- FDCs are infected in the patches and in other lymphoid organs, including the spleen.
- The prions reach the spleen by a B-cell independent route involving complement factors.

# Prion diseases in Animals

- Scrapie
- Transmissible Mink Encephalopathy (TME)
- Chronic Wasting Disease of Mule deer (CWD)
- Bovine Spongiform Encephalopathy (BSE)

# Classic CJD or Creutzfeldt-Jakob disease (human)

- **Symptoms:**
  - Dementia
  - muscle twitching
  - Vision problems

# Scrapie (goats, sheep)

Occurs as infection exclusively in genetically susceptible sheep

There is no evidence of spread to humans

# Scrapie (Symptoms)

- The name scrapie - derived from one of the clinical signs of the condition
- affected flock compulsively scrape off their fleece against rocks, trees, or fences.
- Disease apparently causes an itching sensation in the animals
- Other clinical signs include
  - excessive lip smacking
  - altered gaits
  - convulsive collapse

# BSE or Bovine Spongiform Encephalopathy (cattle)

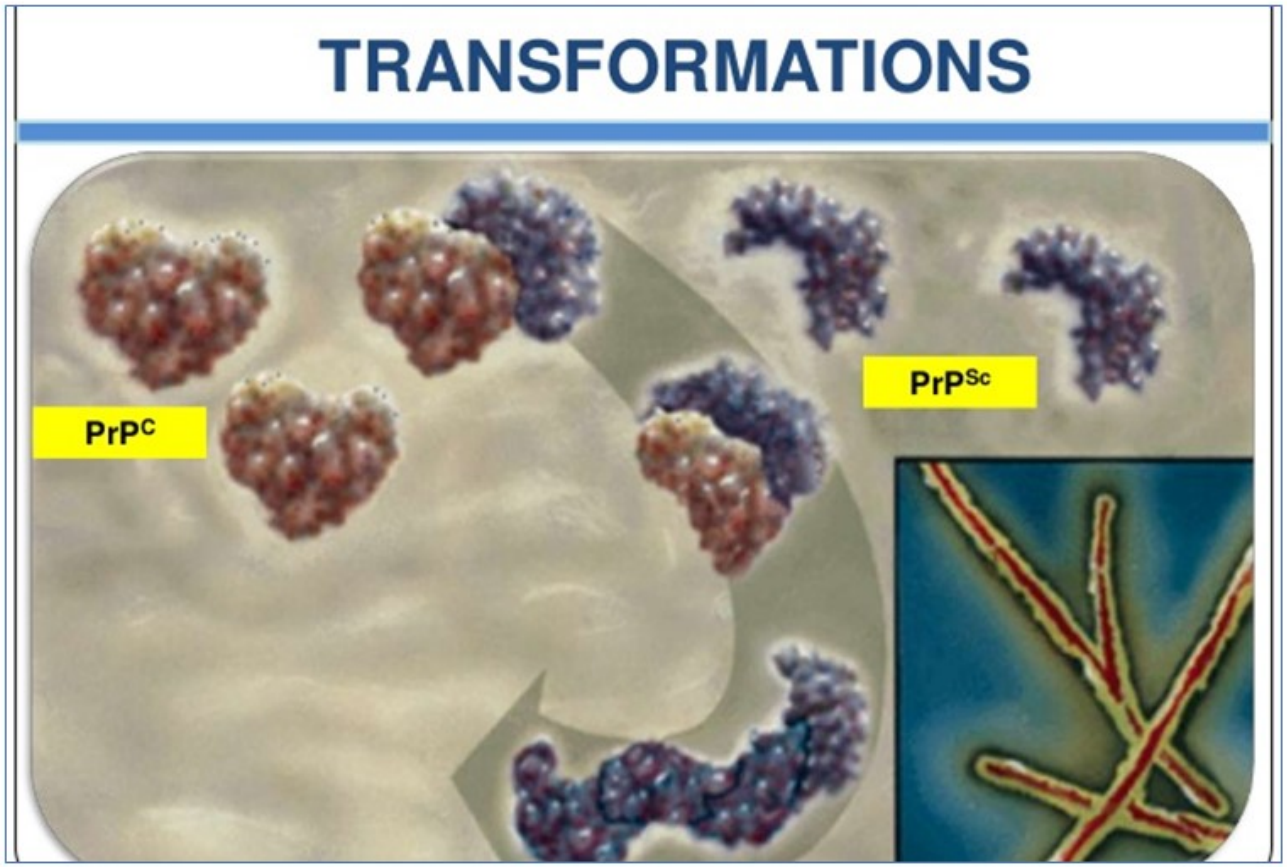
- Also known as "mad cow disease" - because infected animals act strangely and can be aggressive
- Cause for BSE
  - meat and bone meal made from the discarded bones and intestines of slaughtered cows and sheep.
- BSE is a chronic degenerative neurological disease in cows
- part of the brain becomes sponge-like.
- exhibits many different kinds of neurotic symptoms and paralysis,
- eventually death occurs

# BSE - Clinical symptoms

- Extreme sensitivity to external stimuli such as light and sound
- Neurotic changes (depression and nervousness)
- Positional imbalance
- Inability to stand straight or move
- Paralysis in the hind legs
- Paralysis of the whole body before death

# Chronic Wasting Disease (deer, elk)

- Infectious disease in wild deer and elk
- primarily in the western United States
- Drooling, difficulty swallowing, weight loss




Transformation

# Bovine Spongiform Encephalopathy (BSE or mad cow disease)

- Affects cattle;
- Spread is caused by feed containing meat and bone meals from infected cattle and sheep. This practice has been banned in Canada since 2004;
- Can cross the animal-human barrier; and
- The World Organisation for Animal Health (OIE – historically Office international des épizooties) has three risk categories for BSE: negligible risk, controlled risk and undetermined risk. Canada is currently classified as being a controlled risk.

# Chronic Wasting Disease (CWD)

- Affects cervids (deer, elk and moose);
- Spread through animal-to-animal contact and contact with infected material such as soil; and
- So far there is no evidence to suggest CWD can cross the animal-human barrier.

# DIAGNOSIS

## BOVINE SPONGIFORM ENCEPHALOPATHY

# DIAGNOSTIC TEHNIQUES

- Reliable diagnosis of prion disease - through autopsy
  - Tissues of the central nervous system, including the brain and spinal cord, obtained at autopsy are used for diagnosis
- Immunology-based techniques such as
  - Enzyme-linked immunosorbent assay (ELISA)
  - Immunohistochemistry
  - Immunoblotting;
- Confirmatory diagnosis - by performing a bioassay to assess the infectivity of the pathogen
- Histopathological test

# ACKNOWLEDGEMENT


- Part of the presentation viz. photographs , text has been taken from www.google.com. The content is only academic purpose. The contributors are duly acknowledged.


ANY QUESTIONS?
THANKS