Agent causing chronic wasting disease (CWD)

AETIOLOGY

Classification of the causative agent

Chronic wasting disease (CWD) is a contagious prion disease of free-ranging and captive deer, elk, and moose. The cellular prion protein (PrP^C) serves as the normal host-encoded cellular prion protein. It is when PrP^C directly binds to the misfolded isoform PrP^{Sc} that PrP^C adopts the disease-associated conformation. Normal prion proteins can be found most abundantly in the brain and spinal cord.

CWD is a member of the transmissible spongiform encephalopathy (TSE) family of prion diseases, and it is believed there are multiple strains within the United States as well as a strain unique to Norway.

Resistance to physical and chemical action

Temperature:	Highly resistant to heat and radiation (UV, microwave, ionising); inactivation by autoclaving at 134°C (273°F) for 18 minutes at 30 lb/in^2 is suitable, but parameters may vary pending type of sample contaminated.
pH:	Bioavailability of the CWD prion in soil is greater when pH>6.6.
Chemicals/Disinfectants:	Highly resistant to chemical inactivation and few disinfectants effectively inactivate them; primarily, 50% concentrated household bleach with a contact time of 30-60 minutes or sodium hydroxide for 60 minutes are recommended, but concentrations and contact times may vary pending the type of sample contaminated.
Survival:	Remains viable for long periods in fluids, faeces and tissues; persists in soil; partially resistant to protease digestion and can accumulate within neurones, eventually causing neuronic death.

EPIDEMIOLOGY

Hosts

- It is known to affect multiple cervid species including but not limited to: elk (*Cervus canadensis*), moose (*Alces alces*), mule deer (*Odocoileus hemionus*), white-tailed deer (*Odocoileus virinianus*), and reindeer (*Rangifer tarandus*).

Transmission

- Direct:
  - Contact of mucous membranes between affected and healthy animals
- Indirect:
  - Environmental contamination of soil, food, or water
  - Consumption of plants that have uptaken prion proteins from contaminated soil (currently experimentally proven in hamsters only)

Sources

- Blood, tissues, secretions and excretions of sick and dead animals

Occurrence

CWD is present in captive and/or free-ranging deer, elk, and moose. It has been reported in three provinces of Canada, at least 26 states of the continental United States, Norway, Finland, Sweden, and in imported animals in South Korea.

For up-to-date information about the distribution of CWD in North America, see the referenced "Distribution of Chronic Wasting Disease in North America" URL provided by the USGS National Wildlife Health Center. For up-to-date information about the distribution of CWD in Sweden, see the referenced "Map of Chronic Wasting Disease" URL provided by the National Veterinary Institute.

DIAGNOSIS

The minimum incubation period is approximately 16 months, and the average incubation period is approximately 2 to 4 years.

Clinical diagnosis

- CWD is fatal
- Clinical diagnosis may be difficult to observe in free-ranging animals, particularly early in the course of the disease
- Animals may present asymptomatically and die suddenly.
- Cervids can develop progressive weight loss and behavioural changes (lethargy, hyperexcitability, low carriage of head with fixed gaze) over several weeks to months.
- Neurological signs may be subtle but can include: ataxia, head tremors, teeth grinding, and pacing of an enclosure's perimeter
- Animals may develop aspiration pneumonia secondary to oesophageal dilation and/or regurgitation, leading to death
- Polyuria (excessive urination) and polydipsia (excessive drinking) are common signs in the terminal stages

Lesions

- Post-mortem lesions:
    - Nonspecific; often emaciated but some carcases may be in good condition with few or no gross lesions (particularly in the early stages of disease)
    - Megaesophagus and aspiration bronchopneumonia can be seen
    - In summer, patchy retention of winter coat
    - Abomasal or omasal ulcers

Differential diagnoses

- Meningitis, encephalitis, brain abscess
- Fading elk syndrome
- Chronic malignant catarrhal fever

- Chronic epizootic haemorrhagic disease (EHD)
- Starvation, dental attrition
- Nutritional deficiencies, e.g., polioencephalomalacia, copper deficiency
- Toxicities, e.g., ryegrass staggers
- Stress
- Arthritis
- Traumatic Injuries
- Pneumonia

Laboratory diagnosis

Samples

For isolation of agent

- Antemortem:
    - Deer - palatine tonsil
    - Deer, elk - rectal lymphoid tissue
- Post-mortem:
    - Brain (obex), retropharyngeal lymph nodes, tonsils

Serologic Samples

- Serology is not used to make CWD diagnoses

Procedures

Identification of the agent

- Immunohistochemistry is the gold standard for diagnosing CWD
- Histopathology of lymphoid tissues and/or CNS
- Immunoblotting (Western blotting), enzyme-linked immunosorbent assays (ELISA), and rapid antigen-detection strip tests can also be used to screen cervids

Serological tests

- Serology is not feasible for antemortem testing; antibodies are not made against PRP^{Sc}
- Protein misfolding cyclic amplification (PMCA) and real-time quaking-induced conversion assay (RT-QUiC) are newly developed assays that can detect CWD prions at preclinical stages from noninvasive, antemortem samples, but are currently still under development for diagnostic use

PREVENTION AND CONTROL

Sanitary prophylaxis & Control

Captive Herds

- Minimize introduction of outside individuals; maintain a closed herd if possible
    - Replacement animals should be from certified CWD-negative herds
- Utilize strategic fencing to reduce or eliminate contact between captive and wild cervids
- In the United States, mandatory CWD reporting varies by state

Wild Herds

- Control is difficult in free-ranging cervids
- Discouraging congregation of cervids in focal areas can decrease the rate of transmission between animals
    - Behaviors such as feeding or baiting are banned in many areas for this reason
- Many states and provinces restrict transportation of tissues from hunter-killed cervids in endemic areas
- Some areas have culled wild populations of cervids, but efficacy appears to be variable

Medical prophylaxis

- There is no vaccine available for CWD

POTENTIAL IMPACTS OF DISEASE AGENT BEYOND CLINICAL ILLNESS

Risks to public health

- There is currently no evidence that CWD poses a threat to humans, but zoonotic potential is not well understood
- Hunters can have carcasses tested for CWD and should avoid eating meat from animals that are of ill-thrift; gloves should be worn when field-dressing carcasses

Risks to agriculture

- There is currently no evidence that CWD prions have infected any animals other than cervids in endemic areas.
    - Experimentally infected species include: voles, mice, cats, raccoons, squirrel-monkeys
- Decontaminating soil is currently proven to be impractical; soil microorganisms may degrade prions in buried carcasses, and plants are capable of uptaking prion protein and transporting it to aerial tissues (leaves, stems), both of which are currently under investigation for better characterisation.

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The OIE will periodically update the OIE Technical Disease Cards. Please send relevant new references and proposed modifications to the OIE Science Department (scientific.dept@oie.int). Last updated 2019. Written by Marie Bucko and Samantha Gieger with assistance from the USGS National Wildlife Health Center.