

Transmissible Spongiform Encephalopathy (TSE)

DISEASE IN ANIMALS

Transmissible Spongiform Encephalopathies are progressive and fatal neurodegenerative diseases that are believed to be caused by prions. It is known to affect many animal species. A prion is an abnormal, transmissible agent that is able to induce abnormal folding of normal cellular prion proteins in the brain leading to brain damage.

Reporting: Animal TSE cases, including bovine spongiform encephalopathy (BSE), chronic wasting disease (CWD), and scrapie, are reportable in Ohio and all suspected cases must be reported to the Ohio Department of Agriculture (ODA), Division of Animal Industry at (614) 728-6220 or (800) 300-9755 or the USDA APHIS Veterinary Services at (614) 856-4735 or (800) 536-7593.

Transmission: The primary route of transmission is through ingestion of contaminated tissue, primarily from central nervous tissue origin. Scrapie and CWD can be transmitted through direct contact with an infected animal or contaminated environment.

Clinical signs: Infection is typically subclinical for years before onset of clinical signs. Neurologic signs predominate and vary among species; however, all cases are progressive and fatal.

SPECIES	CLINICAL SIGNS
Sheep (scrapie)	Behavioral changes followed by hyperexcitability and high-stepping gait many develop a fixed stare with the head held high, intense pruritus is common. Progresses to ataxia, blindness, convulsions and death.
Goat (scrapie)	Symptoms are variable and may include irritability and loss of inquisitiveness, hyperesthesia, incoordination, posture abnormalities, restlessness, tremors, teeth grinding, salivation, impaired vision or regurgitation of rumen contents.
Cattle (BSE)	Typically, hyperreactive to stimuli, gait abnormalities, and behavioral changes such as aggression, nervousness or frenzy are seen.
Deer/Elk (CWD)	Progressive weight loss is the predominant sign. Progressive behavioral changes, ataxia, head tremors, teeth grinding and hyperexcitability may also be seen. In captive animals perimeter pacing has also been noted.
Cats (FSE)	Uncharacteristic aggression, timidity, gait abnormalities and ataxia initially affecting the hind legs. Some cats develop a rapid, crouching, hypermetric gait and hyperesthesia is common.
Mink (TME)	Early signs can be subtle and may include difficulty eating, swallowing and changes in normal grooming behavior. Later, animals may become hyperexcitable and bite compulsively. Affected mink often carry their tails arched over their backs like squirrels.

Case classification:

- Suspected: a clinical case with signs consistent with a TSE.
- Probable: a clinically suspect case with laboratory evidence from a screening or unvalidated test.
- Confirmed: a case that meets confirmatory testing criteria determined by a state or federal diagnostic laboratory.

DISEASE IN HUMANS

Reporting: Creutzfeldt-Jakob Disease (CJD) is the human form of the disease and cannot be clinically differentiated from other TSEs. Report by the end of the business week any suspected human illness or positive laboratory result to the [local health department](#) (LHD) where the patient resides. If unknown, report to the LHD of the health provider or laboratory.

Human illness: The only TSE known to be zoonotic is BSE or vCJD. The symptoms are very similar to CJD. Sporadic CJD primarily affects persons 50 to 80 years old and has an insidious onset with confusion, poor concentration, lethargy, progressive dementia, intermittent unsteadiness when standing or walking and variable ataxia. Muscle jerks and other neurologic signs appear later. Sporadic CJD progresses rapidly and death usually occurs within three to 12 months (mean seven months). Familial CJD usually has an onset at about 40 years and patients may live for five to 11 years. With vCJD, younger people can be affected and most patients die in six months to two years. The primary route of transmission of vCJD is through ingestion of contaminated tissue.

Personal protection: While BSE is the only known zoonotic TSE, there is still much that is not known about prion diseases. It is recommended to use appropriate PPE in all suspect TSE cases. Standard precautions include the use of protective clothing and the avoidance of penetrating injuries, contamination of abraded skin and ingestion. A BSL-3 lab is and a negative pressure laminar flow hood should be used for tissue manipulations whenever possible. Because prions may be able to survive in the environment for years and are difficult to disinfect, precautions should be taken to avoid contamination of surfaces and equipment.

FOR MORE INFORMATION

Reportable Animal Diseases in Ohio:

[ODA Division of Animal Industry](#)

[OAC Chapter 901:1-21 Dangerously Contagious or Infectious and Reportable Diseases](#)

[USDA Animal and Plant Health Inspection Service](#)

Disease in Animals

[Ohio Animal Disease Diagnostic Laboratory](#)

[USAHA Foreign Animal Diseases "Gray Book"](#)

[Iowa State University Center for Food Security and Public Health Animal Disease Factsheets](#)

[USDA Animal and Plant Health Inspection Service Animal Diseases](#)

[AVMA Public Health Disease Information](#)

Disease in Humans

[ODH Infectious Disease Control Manual \(Creutzfeldt - Jakob disease\)](#)

[CDC Prion Diseases](#)