NVAP Reference Guide: Transmissible Spongiform Encephalopathies

Last Modified:

Transmissible spongiform encephalopathies (TSEs) are rare progressive neurodegenerative disorders that affect both humans and animals and are caused by similar uncharacterized agents that generally produce spongiform changes in the brain. Specific examples of TSEs include classical and Nor98-like scrapie, which affect sheep and goats; bovine spongiform encephalopathy, which affects cattle, and rarely other bovidae (nyala, oryx, eland, kudu, and goats), domestic and exotic cats (feline spongiform encephalopathy), humans (variant Creutzfeldt–Jakob disease), and zoo primates; transmissible mink encephalopathy; and chronic wasting disease of mule deer, white- tailed deer, black-tailed deer, elk and moose.

TSEs are insidious degenerative diseases of the central nervous system. Historically, the diagnosis of TSEs has been based on the occurrence of clinical signs of the disease, which could only be confirmed by postmortem examination of brain tissue using histopathology. More recently, identifying the presence of abnormal prion protein by various techniques has allowed preclinical diagnosis.

A characteristic feature of all TSEs is the lack of a measurable host immune response to the agent, meaning that no antibodies are produced. No conventional serologic test can be used to identify infected animals. Scientists usually diagnose TSEs in the laboratory by immunohistochemistry, western blot, or ELISA performed on brain and or lymphoid tissue which may be followed by one or more supplemental tests.

Three TSEs will be discussed in this chapter; Scrapie, Bovine Spongiform Encephalopathy, and Chronic Wasting Disease.

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