

Mad Cow and Chronic Wasting Disease

Traveler Summary

Key Points

- Mad cow disease (Bovine spongiform encephalopathy [BSE]) is a fatal disease of the brain and nervous system in cattle that is acquired by humans through the consumption of infected beef, causing variant Creutzfeldt-Jakob disease (vCJD).
- Control of the food chain in affected areas is such that risk of vCJD is negligible to the point where it does not need to be considered by travelers.
- Symptoms in humans involve progressive neurological deterioration, including a slow onset of personality changes or psychiatric symptoms, followed by impaired balance or coordination and sudden, involuntary muscle jerks or spasms.
- Consequence of infection is certain death.
- Avoid consuming beef and beef products in any of the few countries where mad cow disease is known to exist; however, this approach is not currently recommended for any destination.
- No vaccine or preventive drugs are available.

Introduction

Mad cow disease is a fatal disease of the brain and nervous system in cattle. The agent that causes mad cow disease, an abnormal protein called a prion, can infect humans after ingestion of beef, causing variant Creutzfeldt-Jakob disease (vCJD), which is a disorder that involves progressive deterioration of the nerve cells in the brain and eventually certain death.

Stricter regulations brought the mad cow disease epidemic in cattle under control in the UK in 1988, and only isolated cases have occurred in animals born since 2000. At the same time, BSE began to appear in other European countries. Stricter regulations and common policies for mad cow disease control and testing were adopted by other EU countries in 2001.

Chronic wasting disease (CWD) also occurs due to another prion affecting the nervous system and has commonly been found in deer, elk, reindeer, and moose in certain areas of North America, Norway, and Finland since 1967. Prevalence is increasing significantly, and deer and moose in 24 states of the US (especially in the Midwest) are now infected. The CWD prion is found in all animal tissues and excreta, including in apparently normal animals; however, no human cases have been reported. Concerned hunters can harvest animals only in known CWD-free zones or have animals tested prior to dressing or eating venison.

Risk Areas

Human cases of vCJD have historically occurred in UK, France, Spain, Ireland, US, Italy, Netherlands, Portugal, Canada, Saudi Arabia, Japan, and Taiwan.

Although countries in the EU have banned the implicated feeding practices since 2001 and have regulations that ban the sale of beef older than 3 years, it is not clear whether any uniformly adequate enforcement or surveillance exists; thus the true extent of the problem is unknown. Nevertheless, BSE in cattle is currently well controlled, occurrence is rare worldwide, and surveillance is generally robust.

Transmission

Cases of vCJD may be transmitted to humans via consumption of beef that has been contaminated with infectious nervous tissue during the slaughtering process.

Direct human-to-human transmission has not been demonstrated, but vCJD may be transmitted through blood transfusion, and possibly through organ transplantation or reuse of medical instruments.

Risk Factors

Risk of acquiring vCJD from eating beef and beef products—even in countries with controlled BSE in cattle—is negligible to the point where it does not need to be considered by travelers. The risk of acquiring vCJD from a blood transfusion has been minimized but not eliminated in the UK, and organ or body tissue transplantation could present a risk.

Symptoms

Symptoms of vCJD involve progressive neurological deterioration, including a slow onset of personality changes or psychiatric symptoms, followed by impaired balance or coordination and sudden, involuntary muscle jerks or spasms.

Consequences of Infection

Death uniformly occurs following symptoms of vCJD.

Need for Medical Assistance

Travelers who have visited countries with known cases of mad cow disease or who develop symptoms of vCJD (including personality changes) should seek medical consultation.

Prevention

Nonvaccine

Avoid consumption of all beef and beef products in countries where mad cow disease is known to exist to eliminate any risk of acquiring vCJD from food; however, this approach is not currently recommended for any destination due to the lack of measurable risk to travelers. Milk and milk products do not pose a risk for vCJD.

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