CONDITION

# Bovine spongiform encephalopathy

Learn what bovine spongiform encephalopathy is, how it relates to variant Creutzfeldt-Jakob disease in people, and when to seek medical advice.

**WHAT IT IS**
Rare fatal human prion brain disease linked to BSE

**BODY SYSTEM**
Nervous system

**MAIN SYMPTOMS**
Behavioral changes, painful sensations, memory problems, poor balance

**WHAT CAUSES IT**
Abnormal prions from contaminated food

**HOW IT IS DIAGNOSED**
Specialized examination of brain tissue

**MAIN TREATMENT**
Supportive and palliative care

**CAN IT BE CURED?**
No, no treatment stops brain damage

**IS IT AN EMERGENCY?**
Sometimes, choking or a prolonged seizure

**LONG-TERM OUTLOOK**
Progressive loss of brain function and death

## Overview

Bovine spongiform encephalopathy, or BSE, is a progressive and fatal brain disease that affects cattle. It is commonly called "mad cow disease." BSE belongs to a group of rare conditions called transmissible spongiform encephalopathies, or prion diseases.

Prions are proteins that have folded into an abnormal shape. Unlike bacteria and viruses, they do not contain genetic material. An abnormal prion can cause normal proteins in the brain to change shape as well. Over time, this damages brain tissue and creates a sponge-like appearance under a microscope.

People do not develop BSE itself. However, exposure to food contaminated with the agent that causes classical BSE has been linked to a rare human prion disease called variant Creutzfeldt-Jakob disease, or vCJD.

Variant CJD is different from sporadic Creutzfeldt-Jakob disease, which usually occurs without a known cause and is not linked to eating beef. It also differs from inherited prion diseases and rare cases caused by certain medical exposures.

BSE matters because prion diseases are fatal and there is no treatment that can stop or reverse them. Food-safety, animal-feed, cattle-surveillance, and tissue-removal rules are designed to prevent infected material from entering the food supply. These controls have greatly reduced the opportunity for human exposure.

## Symptoms

Symptoms in cattle

Cattle with BSE may develop gradual changes in behavior and movement. Possible signs include nervousness, unusual reactions to touch or sound, difficulty standing, poor coordination, and weight loss despite continued appetite. These signs are not specific to BSE, so veterinary and laboratory assessment is necessary.

People who notice concerning neurologic signs in cattle should contact a veterinarian or the appropriate animal-health authority. They should not attempt to diagnose the animal themselves.

Symptoms of variant CJD in people

Variant CJD usually begins with symptoms that can resemble more common medical or mental health conditions. Early symptoms may include:

As the illness progresses, people may develop worsening coordination, confusion, dementia, speech problems, muscle stiffness, vision changes, and loss of independence. Swallowing and movement may eventually become difficult.

These symptoms do not automatically mean someone has vCJD. Depression, tingling, memory concerns, and balance problems have many other possible causes. A history of eating beef, including beef in a country that experienced BSE, is also not enough to establish a diagnosis.

Arrange a medical appointment for persistent or progressive changes in thinking, behavior, sensation, balance, or movement. Emergency care is appropriate for sudden one-sided weakness, a sudden severe headache, new inability to speak, severe breathing difficulty, choking, loss of consciousness, or a prolonged seizure. Sudden symptoms are more likely to have another cause, such as stroke, but still require prompt care.

- Depression, anxiety, withdrawal, or other behavioral changes
- Unusual painful sensations, such as burning or tingling

- Problems with memory, thinking, or concentration
- Difficulty walking or poor balance
- Involuntary movements or muscle jerks

## Causes and risk factors

Classical BSE is thought to have spread among cattle through feed containing contaminated tissues from infected ruminants. Ruminants are animals such as cattle, sheep, and goats. Feed restrictions were introduced to interrupt this route of transmission.

Atypical BSE is a separate form detected mainly in older cattle. It appears to arise spontaneously at a very low level rather than through contaminated feed. Surveillance programs test appropriate cattle and help keep high-risk tissues out of human and animal food chains.

Variant CJD in people has been linked to consuming food contaminated with tissues from cattle affected by classical BSE. Tissues from the brain, spinal cord, and certain other parts of an infected animal carry the greatest concern. Modern food-safety controls are intended to remove these materials.

BSE is not spread through ordinary contact with cattle or with a person who has vCJD. It is not known to spread by coughing, sneezing, touching, sexual contact, or sharing household items. Milk and dairy products have not been shown to transmit BSE.

Cooking does not reliably destroy prions. Prions also resist some routine cleaning and sterilization methods. Healthcare facilities therefore use special procedures when instruments may have contacted high-risk tissue from someone with known or suspected prion disease.

A small number of vCJD transmissions through blood transfusion have been reported. Blood-collection services use donor-screening and safety policies to reduce this risk. Eligibility rules can change as evidence and blood-supply needs change, so people with questions about donation should ask the relevant blood service.

The main recognized risk factor for vCJD is past exposure to food contaminated during a BSE outbreak. Because symptoms may not appear for years, a clinician assessing possible prion disease may ask where a person lived or traveled and whether they received blood transfusions or certain medical procedures.

## Diagnosis

There is no routine screening test that can tell a healthy person whether they were exposed to BSE or will develop vCJD. Testing is not generally performed based only on past residence, travel, or beef consumption.

When someone has progressive neurologic or psychiatric symptoms, a clinician begins with a detailed history and examination. The history may cover the timing of symptoms, travel and residence, family medical history, medications, substance use, blood transfusions, and past operations. A neurologic examination checks memory, speech, eye movements, strength, sensation, balance, coordination, and reflexes.

The evaluation also looks for more common and potentially treatable conditions. Blood tests may help identify metabolic problems, vitamin deficiencies, infection, autoimmune disease, or medication effects. The exact tests depend on the person's symptoms.

Magnetic resonance imaging, or MRI, creates detailed pictures of the brain. Certain MRI patterns can support suspicion for vCJD, but an MRI alone cannot confirm the diagnosis. An electroencephalogram, or EEG, may be used to record the brain's electrical activity. Testing of cerebrospinal fluid-the fluid surrounding the brain and spinal cord-may help evaluate prion disease and rule out other causes.

Specialized prion tests are interpreted together with the symptoms, examination, and imaging. In selected cases, testing tonsil tissue may provide useful evidence because abnormal prion protein can collect there in vCJD. This is not a routine screening procedure.

A definite diagnosis generally requires examination of brain tissue using specialized laboratory methods, often after death. Brain biopsy during life is uncommon because it carries risks and may not change treatment. It may be considered only when doctors need to investigate another potentially treatable condition.

Because diagnosis is complex, suspected cases are usually assessed by a neurologist and a specialist center familiar with prion diseases. Public-health authorities may also be notified according to local requirements.

## Treatment and management

There is no approved treatment that cures vCJD or stops the underlying prion-related brain damage. Management focuses on comfort, safety, symptom relief, and support for the person and family.

A care plan may involve neurology, primary care, nursing, rehabilitation, palliative care, mental health services, nutrition specialists, and social work. Palliative care is specialized care that aims to reduce symptoms and improve quality of life. It can begin alongside other medical

evaluation and is not limited to the final stage of illness.

Treatment may address anxiety, depression, pain, muscle stiffness, involuntary movements, sleep problems, or seizures. The choice of medicine depends on the person's symptoms, overall health, and risk of side effects. Medicines do not remove prions, and treatment should be individualized by the clinical team.

Physical and occupational therapists may suggest ways to reduce falls and make daily activities safer. Mobility aids, home modifications, and help with bathing or dressing may become necessary. Speech and language therapists can assess communication and swallowing.

If swallowing becomes difficult, the care team may recommend changes in food texture, eating position, or feeding support. These decisions should consider comfort, aspiration risk, the person's wishes, and the expected benefits and burdens of each option.

For cattle, there is no treatment for BSE. Suspected animals are managed through veterinary and public-health systems. Affected cattle do not enter the food supply.

## Complications

Variant CJD causes continuing loss of brain function. The pace and pattern can vary, but affected people eventually need extensive help with daily life.

Possible complications include falls, injuries, difficulty communicating, loss of mobility, and inability to make independent decisions. Swallowing problems may allow food or fluid to enter the lungs. This is called aspiration and can lead to pneumonia. Reduced movement can contribute to pressure injuries, blood clots, constipation, and infections.

Behavioral and cognitive changes may be distressing for both the affected person and those close to them. Caregivers may experience exhaustion, grief, financial strain, or difficulty coordinating services. Respite care, social work support, counseling, and prion-disease organizations can help families plan and cope.

Prion diseases are fatal. Early conversations about goals of care can help the healthcare team understand what matters most to the person. These discussions may include preferred care settings, emergency treatment, feeding support, symptom control, and who should make decisions if the person loses decision-making capacity.

## Follow up

A person who is well but worried about a possible past food exposure does not usually need emergency care. A routine appointment with a primary care clinician can help review the exposure and explain whether any specific follow-up is appropriate. There is currently no preventive medicine or standard test for an asymptomatic person after dietary exposure.

People with ongoing or worsening neurologic symptoms should arrange medical assessment rather than assuming the cause is vCJD. Referral to neurology may be appropriate. Bringing a written symptom timeline, travel and residence history, medication list, and details of past transfusions or procedures can help the evaluation.

For someone being assessed for or living with suspected vCJD, follow-up should address changing symptoms, falls, swallowing, nutrition, communication, caregiver needs, and advance care planning. The team may also explain any precautions needed for surgery, tissue handling, or medical instruments. Routine social contact does not require isolation.

Seek urgent or emergency help for serious breathing or choking problems, a prolonged seizure, a major injury, loss of consciousness, or sudden stroke-like symptoms. For gradual changes, contact the regular clinical team promptly so that symptoms and support needs can be reviewed.

## Sources

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