Mad Cow Disease, BSE
Creutzfeldt-Jakob Disease

Mad cow disease is a brain disorder that occurs in cattle. The clinical name for mad cow disease is bovine spongiform encephalopathy or BSE for short. It is caused by a twisted protein called a *prion.* Mad cow disease is a fatal condition that affects the nervous system of cattle. In an animal with mad cow disease, the brain develops tiny holes, the animal loses control over its movements, its behavior changes, and the animal seems to go mad. Scientists think BSE may have come from a similar disease in sheep, called *scrapie*. The tiny holes in the brain are areas where brain cells have died. Because of these holes, the brain of an animal with mad cow disease looks like a sponge. This is the reason for the clinical name of the disease, bovine spongiform encephalopathy.

The connection between BSE and humans was uncovered in Great Britain in the 1990s when several young people died of a rare brain disorder that usually strikes the elderly. The brain disorder is called **Creutzfeldt-Jakob Disease** (CJD). The new version was called **new variant Creutzfeldt-Jakob Disease** (nvCJD) because it began to affect young people.

All of these similar disorders of the brain seem to be caused by *prions* which are twisted proteins. As far as we can tell, these twisted proteins are not needed. We do not know what purpose normal, untwisted prion proteins serve. Cells tend to shove prions into vesicles and the prions build up in the vesicles. The prions eventually burst out of the vesicles. This kills the cell. After this, the released prions probably enter the surrounding cells and repeat the process.

SYMPTOMS

Symptoms (problems) in animals include an excitable or nervous temperament to simple things, such as touching the skin. Another symptom is a progressive unsteadiness of gait. This makes the animal unable to stand up. So far, the disease is 100% fatal.

The human equivalent of Mad Cow Disease is Cruetzfeldt-Jakob Disease, or CJD. Symptoms of CJD include memory loss and emotional instability that includes inappropriate outbursts. Other symptoms include an unsteady gait that progresses to severe weakness and severe *dementia* (brain is not thinking right) that gets worse and worse. The patient usually dies within a year after the first symptoms appear.

TREATMENT

The disease is currently untreatable and incurable. It is fatal.

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