Q&A: Creutzfeldt-Jakob Disease
And Mad Cow Disease

1 WHAT IS CREUTZFELDT-JAKOB DISEASE?

Creutzfeldt-Jakob disease (CJD) is one of a group of rare diseases called "transmissible spongiform encephalopathies (TSEs)." These diseases affect people and some kinds of animals. They attack the central nervous system and invade the brain, causing damage and eventual death.

Creutzfeldt-Jakob disease is very rare. Only about 1 case per million people occurs each year worldwide.

2 IS CREUTZFELDT-JAKOB DISEASE THE SAME AS 'MAD COW DISEASE?'

They are related, but not the same. Both are TSEs. But only *people* get Creutzfeldt-Jakob disease, and only *cattle* get mad cow disease (bovine spongiform encephalopathy, or BSE).

Cattle are thought to get mad cow disease when they are fed contaminated beef by-products or bone meal.

It is possible for people to get a form of Creutzfeldt-Jakob disease (variant CJD, or vCJD) by eating beef from cattle with mad cow disease. The likelihood is very small, but not zero. Almost all of the people who have been infected this way ate beef while living in England during the 1980s and 1990s.

3 WHAT CAUSES CREUTZFELDT-JAKOB DISEASE?

CJD is caused by a protein called a *prion*. (It is *not* caused by germs like bacteria and viruses.) Prions may cause other proteins in the brain to change shape, killing brain cells and making areas of the brain look spongy under a microscope. Most of the time, we don't know how or why this happens.

There are 4 forms of CJD:

- **Sporadic.** 85% to 90% of CJD cases are sporadic: the cause is unknown. Most sporadic cases occur in people over 50. These people usually die within months. Sporadic CJD is not related to mad cow disease.
- **Familial.** 5% to 10% of CJD cases are inherited. People usually develop familial CJD at an earlier age than the sporadic form, and the course of the illness is longer. Familial CJD is not related to mad cow disease.
- **Acquired.** About 1% of CJD cases have been caused by contamination with tissue from an infected person. This usually has occurred through medical procedures such as dura mater (covering of spinal cord) transplants or through the use of contaminated surgical instruments. Today, no transplants are taken from people known to be infected with CJD, and surgical instruments used on people with CJD are never used again.
- **Variant (vCJD).** First reported in 1996, this form of CJD appears to be caused by eating beef from cattle infected with mad cow disease. It is very rare.

4 DOES CREUTZFELDT-JAKOB DISEASE SPREAD FROM PERSON TO PERSON?

There is some evidence that vCJD may be transmitted by blood transfusion. The general public does not need to do anything special to protect against CJD. All other forms of CJD are NOT known to be spread from person to person through the air, through blood, or through sexual contact.

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Health care workers and others who may be exposed to the blood or other body fluids of a CJD patient should take standard precautions.

5 WHAT ARE THE SYMPTOMS OF CREUTZFELDT-JAKOB DISEASE?

Symptoms may occur months or even years after exposure. Early symptoms include loss of memory, clumsiness, slurred speech, and visual problems or hallucinations. As the illness gets worse, so do mental problems. A person may also have muscle twitches, weakness of the hands, feet, and limbs, and eventually coma.

6 HOW IS IT DIAGNOSED?

Doctors sometimes test for sporadic CJD by testing electrical activity in the brain and examining spinal fluid. Familial CJD may be diagnosed with a blood test for the abnormal gene.

Diagnosis can be confirmed only through brain biopsy or autopsy (an examination of the brain after death). Autopsies are important in helping researchers track CJD.

7 HOW CAN CJD BE PREVENTED?

One way CJD can be prevented is by stopping the spread of mad cow disease. Since 1990, the U.S. Department of Agriculture (USDA) has been testing American cattle and beef for BSE.

An expanded BSE screening program was started in June, 2004, after a cow slaughtered in Washington State tested positive for the disease. Since then, more than 650,000 animals have been tested – about 1 of every 90 cows. A second BSE-positive cow was found in 2005 and a third in 2006. No other cases of BSE have been seen in other cattle.

Import restrictions. In 1989, the USDA strictly limited the importation of cattle, sheep, and goats, and certain products from these animals, from countries where BSE was known to exist. These restrictions were later extended to include certain animals and products from all European countries.

A **feed ban** in place since 1997 forbids feeding U.S. livestock any cattle meat or bone meal that has been broken down by a process called "rendering." The Harvard Center for Risk Assessment concluded that this rule provides a major defense against BSE and CJD.

8 CAN CJD BE TREATED?

So far, there is no cure for CJD. The disease always results in death, usually within a year after symptoms start, but sometimes within months or even weeks.

Treatment can ease symptoms and discomfort – for instance, pain medicines and frequent turning of the patient to avoid bedsores.

Researchers are experimenting with drug therapies and studying the use of antibodies to prevent and treat CJD and other prion diseases. They are also trying to make a vaccine.

9 WHERE CAN CJD FAMILIES FIND SUPPORT?

For support and information, call the CJD Foundation Help Line (1-800-659-1991) or visit **www.cjdfoundation.org**

For help in arranging an autopsy, call the National Prion Disease Pathology Surveillance Center (216-368-0587). Or call **311** and ask for the CJD Surveillance Coordinator at the New York City Department of Health and Mental Hygiene.

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