Fact Sheet

Transmissible Spongiform Encephalopathies

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What is Transmissible Spongiform Encephalopathies?

Transmissible spongiform encephalopathies (TSEs), also known as prion diseases, are a group of rare degenerative brain disorders characterized by tiny holes that give the brain a "spongy" appearance. These holes can be seen when brain tissue is viewed under a microscope.

- Creutzfeldt-Jakob Disease (CJD) is the most well-known of the human TSEs. It is a rare type of dementia that affects about one in every one million people each year.
- Other human TSEs include kuru, fatal familial insomnia (FFI), and Gerstmann-Straussler-Scheinker disease (GSS).
- Kuru was identified in people of an isolated tribe in Papua New Guinea and has now almost disappeared. FFI and GSS are extremely rare hereditary diseases, found in just a few families around the world.
- A new type of CJD, called variant CJD (vCJD), was first described in 1996 and has been found in Great Britain and several other European countries. The initial symptoms of vCJD are different from those of classic CJD and the disorder typically occurs in younger patients. Research suggests that vCJD may have resulted from human consumption of beef from cattle with a TSE disease called bovine spongiform encephalopathy (BSE), also known as "mad cow disease."
- Other TSEs found in animals include scrapie, which affects sheep and goats; chronic wasting disease, which affects elk and deer; and transmissible mink encephalopathy. In a few rare cases, TSEs have occurred in other mammals such as zoo animals. These cases are probably caused by contaminated feed. CJD and other TSEs also can be transmitted experimentally to mice and other animals in the laboratory.

How is it Spread?

Human TSEs can occur three ways: *sporadically*; as *hereditary* diseases; or through *transmission* from infected individuals. Sporadic TSEs may develop because some of a person's normal prions spontaneously change into the infectious form of the protein and then alter the prions in other cells in a chain reaction. Inherited cases arise from a change, or mutation, in the prion protein gene that causes the prions to be shaped in an abnormal way. This genetic change may be transmitted to an individual's offspring. Transmission of TSEs from infected individuals is relatively rare. TSEs cannot be transmitted through the air or through touching or most other forms of casual contact. However, they may be transmitted through contact with infected tissue, body fluids, or contaminated medical instruments. Normal sterilization procedures such as boiling or irradiating materials do not prevent transmission of TSEs.

Signs and Symptoms

Symptoms of TSEs vary, but they commonly include personality changes, psychiatric problems such as depression, lack of coordination, and/or an unsteady gait. Patients also may experience involuntary jerking movements called myoclonus, unusual sensations, insomnia, confusion, or memory problems. In the later stages of the disease, patients have severe mental impairment and lose the ability to move or speak.

Diagnosis

TSEs can only be confirmed by taking a sample of brain tissue during a biopsy or after death. Doctors, however, can do a number of tests to help diagnose prion diseases such as CJD, or to rule out other diseases with similar symptoms.

These tests include:

- MRI scans of the brain
- Samples of fluid from the spinal cord (spinal tap)
- Electroencephalogram, which analyzes brain waves; this painless test requires placing electrodes on the scalp
- Blood tests
- Neurologic and visual examinations to evaluate for nerve damage and vision loss

Treatment

There is currently no treatment that can halt progression of any of the TSEs. Treatment is aimed at alleviating symptoms and making the patient as comfortable as possible. TSEs tend to progress rapidly and usually culminate in death over the course of a few months to a few years.

For more sources of information on this topic visit:
ST. CLAIR COUNTY HEALTH DEPARTMENT www.scchealth.co
MICHIGAN DEPARTMENT OF HEALTH AND HUMAN SERVICES www.michigan.gov/mdhhs
CENTERS FOR DISEASE CONTROL AND PREVENTION www.cdc.gov
NATIONAL INSTITUTE OF NEUROLOGICAL DISORDERS & STROKE www.ninds.nih.gov