Georgia Department of Public Health

Peachtree Street NW, 15th Floor
Atlanta, Georgia 30303-3142
www.health.state.ga.us

CREUTZFELDT-JAKOB DISEASE FACT SHEET

(Jakob-Creutzfeldt syndrome, Subacute spongiform encephalopathy, CJD)

Agent: CJD is caused by a filterable, self-replicating agent called a prion. Prions are distinct from viruses in not containing nucleic acids (DNA or RNA). Infection with a prion causes certain host proteins to fold abnormally resulting in nervous system disease.

Brief Description: Creutzfeldt-Jakob Disease (CJD) is a rare and fatal brain disorder. Most cases of classic CJD are sporadic and the source of the disease is not known. Some cases of CJD are inherited genetically. Iatrogenic CJD cases can occur as a result of contamination with nervous system tissue, cornea grafts or pituitary gland-derived growth hormone from an infected person. Variant CJD (vCJD) is caused by exposure to bovine spongiform encephalopathy (BSE), or "mad cow disease," a prion disease found in cattle. Initial symptoms of CJD may include: depression, memory lapses, dementia, unsteadiness, and lack of coordination. Later, symptoms may progress to jerky movements, rigid limbs, blindness and incontinence. Eventually, a person with CJD loses the ability to move or speak.

Reservoir: Humans are the only known reservoir for CJD.

Mode of Transmission: The mode of transmission for most cases is not known. In cases of iatrogenic CJD, transmission has occurred during surgery by the reuse of contaminated surgical instruments, transplant of infected corneas or by treatment with human growth hormone that is derived from human pituitary glands of someone with CJD. Recent cases in the UK have also resulted from the receipt of transfused blood contaminated with the vCJD agent.

Incubation period: The incubation period can last 15 months up to 30 years. Once symptoms begin, the disease is rapidly progressive, generally resulting in death within two years.

Laboratory Criteria for Diagnosis:

Diagnostic Testing (all testing is done at the National Prion Disease Pathology Surveillance Center at Case Western Reserve University).

1. Specimen needed:
    A. Brain tissue: 1-5 grams of brain tissue collected during autopsy
    B. Brain tissue: 0.01-0.5 grams of brain tissue collected during biopsy
    C. Cerebrospinal Fluid (CSF): 2-5 ml of CSF collected by lumbar puncture
2. Specimen preparation: Freeze at –70°C and ship on dry ice.
3. Lab Form:
    A. Brain tissue (autopsy): Autopsy Information Sheet
    B. Brain tissue (biopsy): Biopsy Information Sheet
    C. CSF: CSF Test Request Form

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4. Lab Test Performed:
   In unfixed brain tissue: Presence of protease-resistant or scrapie protein (PrP)
   In fixed brain tissue: Microscopic examination
   In CSF: Presence of 14-3-3 protein
5. Laboratory: Submit specimens to the National Prion Disease Pathology Surveillance Center at the Division of Neuropathology of Case Western Reserve University

Comment: Visit http://case.edu/medicine/pathology/divisions/prion-center/ for more specific information on laboratory procedure and associated forms.

Period of Communicability: Central nervous system tissues are infective throughout symptomatic illness and post-mortem. Infectivity during incubation period is not known.

Treatment: There is currently no known treatment for CJD. There are drugs that may relieve symptoms and make the patient more comfortable.

Investigation and Follow-Up: Investigate cases to determine the possible source of infection. Obtain a complete medical history, including surgical and dental procedures, exposure to human hormones or transplanted tissue, travel history and family history of dementia.

Reporting: While CJD is not a highly communicable disease, surveillance for vCJD) is needed to ensure risk containment for bovine spongiform encephalopathy (BSE) in the food supply. For this reason, Georgia mandates reporting of suspect and confirmed CJD cases under the age of 55. Timely notification will allow for gathering of background information to determine whether the case is likely to be classic or variant CJD. Timely action is also needed to discuss the importance of biopsy and/or autopsy confirmation of atypical CJD cases.

Report probable, suspected, or definite cases among persons less than 55 years of age **WITHIN 7 DAYS** electronically through the State Electronic Notifiable Disease Surveillance System (SENDSS) at http://sendss.state.ga.us, or complete and mail a GA Notifiable Disease Report Form (#3095). Complete GDPH Form "CJD Case Report Form." *****

IMMEDIATELY report any cluster of Creutzfeldt-Jakob Disease by telephone to the local health department, District Health Office, or the Epidemiology Branch at 404-657-2588. If calling after hours, report cases to the Epidemiology Branch answering service at 770-578-4104.

***** The CJD case report form is a complex one, reflecting the complex clinical syndrome of the disease. GDPH urges those who will be reporting and investigating these cases to seek the assistance of the patient's physician (neurologist, pathologist, attending physician) when completing the case report form. GDPH epidemiologists are also available to facilitate the case investigation and completion of the case report form.

References:

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1. Centers for Disease Control and Prevention. Bovine Spongiform Encephalopathy and Creutzfeldt-Jakob Disease at http://www.cdc.gov/ncidod/dvrd/vcjd/qa.htm
2. Chin J, ed. Encephalopathy, Subacute Spongiform. In: Control of Communicable Diseases Manual. 17th ed. Washington, DC: American Public Health Association, 2000: 183-186.
3. Centers for Disease Control and Prevention. Epidemiology of vCJD and BSE at https://www.cdc.gov/prions/vcjd/index.html

Links:

- CDC Creutzfeldt-Jakob Disease https://www.cdc.gov/prions/cjd/index.html
- National Prion Disease Pathology Surveillance Center http://www.cjdsurveillance.com/
- The National Creutzfeldt-Jakob Disease Surveillance Unit (UK site) http://www.cjd.ed.ac.uk/
- Creutzfeldt-Jakob Disease Foundation http://www.cjdfoundation.org/