A Brief Primer on Creutzfeldt-Jakob Disease

Creutzfeldt-Jakob Disease (CJD) is a neurodegenerative illness and the most common of the human prion diseases. The causative agent of CJD is believed to be a prion - a proteinaceous infectious particle. Prions are thought to induce abnormal folding of cellular proteins in the brain, leading to brain damage and the characteristic signs and symptoms of the disease (1). Prion diseases are usually rapidly progressive and always fatal (2).

CJD Reporting
While CJD is not a communicable disease, except under extraordinary circumstances, surveillance for variant CJD (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 persons under the age of 55. Timely notification will allow 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. In conjunction with the National Prion Disease Pathology Surveillance Center at Case Western University in Cleveland, Ohio, the Georgia Division of Public Health (GDPH) can arrange for testing and/or autopsies to be performed on persons presumed to have died from CJD (of any type). The National Prion Disease Pathology Surveillance Center will help establish the diagnosis of prion disease by analyzing cerebrospinal fluid (CSF), blood, and brain tissue obtained either at biopsy or autopsy and will identify the precise type of prion disease (sporadic, familial, or acquired, defined below) by examining the prion protein and the prion protein gene, once the diagnosis of prion disease has been established (6).

GDPH has developed a form in the State Electronic Notifiable Disease Surveillance System (SendSS) where suspect and/or confirmed cases of CJD (of any type) can be reported. Although mandatory reporting requests apply to suspect/confirmed cases under the age of 55 years old, GDPH does, however, support reporting of any and all suspect/confirmed cases regardless of patient age.

The SendSS form for reporting CJD 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 SendSS form. GDPH epidemiologists are also available to facilitate the case investigation and completion of the SendSS form.

A Brief Primer on Creutzfeldt-Jakob Disease
There are three types of CJD: sporadic, familial, and acquired. Approximately 85% of CJD cases are sporadic. This sporadic disease occurs worldwide, at a rate of approximately one case per 1 million population per year, although rates of up to two cases per million are not unusual (2). The risk of sporadic CJD increases with age, and in persons over 50 years of age, the annual rate is approximately 3.4 cases per million (2). Most sporadic CJD patients develop a rapidly progressing dementia, often accompanied by involuntary muscle spasms, resulting in death within months of the first clinical symptoms. Other initial signs related to illness include ataxia and sight problems. For some the disease duration can be longer than two years (3). In recent years, the United States has reported fewer than 300 cases of sporadic CJD per year (2).

Whereas the majority of cases of CJD occur as sporadic disease, a smaller proportion of patients (5-15%) develop familial CJD because of inherited mutations of the prion protein gene (2). The remainder of CJD cases are acquired either through human-to-human transmission e.g., through use of contaminated surgical instruments, tissue implants, or use of human hormones extracted from the organs of CJD-affected human cadavers, or in the case of vCJD, through the ingestion of prion-contaminated meat.

Variant CJD is a rare, degenerative, fatal brain disorder in humans. Although experience with this new disease is limited, evidence to date indicates vCJD has never been transmitted through direct contact of one person with another. However, a case of probable transmission of vCJD through transfusion of blood components from an asymptomatic donor who subsequently developed the disease has been reported (4).

As of November 2005, a total of 185 cases of vCJD have been reported from 11 countries: 158 from the United Kingdom, 15 from France, 3 from Ireland, 2 from the United States, and 1 each from Canada, Italy, Japan, the Netherlands, Portugal, Saudi Arabia, and Spain (note: the Canadian, one of the Irish, Japanese and U.S. cases were reported in persons who visited or resided in the United Kingdom during a key exposure period of the U.K. population to the BSE agent) (4). Variant CJD has never been reported in a person who did not have a history of exposure within a country where the cattle disease, BSE or “mad cow disease”, was occurring (4).

Persons who develop vCJD become infected through consumption of cattle products contaminated with the agent of BSE (4). The molecular similarity between the bovine and human prion provides strong evidence that vCJD has been acquired from cattle affected by BSE, which occurred in epidemic proportions in the United Kingdom (with limited spread to other countries) in the
1980’s. Variant CJD has well defined and consistent clinical and pathological features that make it relatively easy to identify and distinguish from classic CJD (5). In contrast to classic CJD, vCJD as witnessed in the United Kingdom predominantly affects younger people, has atypical clinical features, with prominent psychiatric or sensory symptoms at the time of clinical presentation and delayed onset of neurologic abnormalities. These neurologic abnormalities include ataxia within weeks or months and dementia and myoclonus late in the illness. Variant CJD also presents as an illness with a duration of at least 6 months and a diffusely abnormal non-diagnostic electroencephalogram (2).

Classic CJD characteristics, as compared to variant CJD, are presented in the table below.

### Clinical and Pathologic Characteristics Distinguishing Classic CJD from Variant CJD

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Classic CJD</th>
<th>Variant CJD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Median age at death</td>
<td>68 years</td>
<td>28 years</td>
</tr>
<tr>
<td>Median duration of illness</td>
<td>4-5 months</td>
<td>13-14 months</td>
</tr>
<tr>
<td>Clinical signs and symptoms</td>
<td>Dementia; early neurologic signs</td>
<td>Prominent psychiatric/behavioral symptoms; painful dyesthesias; delayed neurologic signs</td>
</tr>
<tr>
<td>Periodic sharp waves on electroencephalogram</td>
<td>Often present</td>
<td>Often absent</td>
</tr>
<tr>
<td>“Pulvinar sign” on MRI*</td>
<td>Not reported</td>
<td>Present in &gt;75% of cases</td>
</tr>
<tr>
<td>Presence of “florid plaques” on neuropathology</td>
<td>Rare or absent</td>
<td>Present in large numbers</td>
</tr>
<tr>
<td>Immunohistochemical analysis of brain tissue</td>
<td>Variable accumulation</td>
<td>Marked accumulation of protease-resistance prion protein</td>
</tr>
<tr>
<td>Presence of agent in lymphoid tissue</td>
<td>Not readily detected</td>
<td>Readily detected</td>
</tr>
<tr>
<td>Increased glycoform ratio on immunoblot analysis of protease-resistance prion protein</td>
<td>Not reported</td>
<td>Marked accumulation of protease-resistance prion protein</td>
</tr>
</tbody>
</table>


*An abnormal signal in the posterior thalami on T2- and diffusion-weighted images and fluid-attenuated inversion recovery sequences on brain magnetic resonance imaging (MRI); in the appropriate clinical context, this signal is highly specific for vCJD.

Content source: National Center for Infectious Diseases http://www.cdc.gov/ncidod/dvrd/cjd/index.htm

Please contact Meghan M. Weems, M.P.H. (mmweems@dhr.state.ga.us or 404-657-6442) with any difficulties you may have when investigating or reporting suspect/confirmed cases of CJD.

This article written by Meghan M. Weems, M.P.H.

**References**


2. CDC: Index page: CJD. Retrieved February 08, 2006 from the Centers for Disease Control and Prevention website: http://www.cdc.gov/ncidod/dvrd/cjd/


Breast cancer is the most commonly diagnosed cancer among Georgia women.

**BREAST CANCER**

- Breast cancer is most commonly diagnosed cancer among Georgia women.
- Breast cancer accounts for 32% of all new cancer cases among women.
- Over 5,600 new cases of breast cancer will be diagnosed in Georgia in 2005.
- One in eight American women will develop breast cancer in her lifetime.
- White women are more likely to be diagnosed with breast cancer than black women, but black women are more likely to die from the disease.

**RISK FACTORS**

- Increasing age
- Personal or family history
- White race
- A long menstrual history
- Never having children or having first child after age 30
- Recent use of oral contraceptives or postmenopausal estrogens
- Previous breast radiation
- Consuming two or more drinks of alcohol daily
- Obesity
- Physical inactivity

**PREVENTION**

The best strategy is to avoid the modifiable risk factors: excessive alcohol, obesity, and physical inactivity.

**Data source:** Georgia Comprehensive Cancer Registry (1999-2002)

**Date updated:** December 2005

**Publication number:** DPHOS.115H

The cumulative numbers in the above table reflect the date the disease was first diagnosed rather than the date the report was received at the state office, and therefore are subject to change over time due to late reporting. The 3 month delay in the disease profile for a given month is designed to minimize any changes that may occur. This method of summarizing data is expected to provide a better overall measure of disease trends and patterns in Georgia.

* Other syphilis includes latent (unknown duration), late latent, late with symptomatic manifestations, and neurosyphilis.

### AIDS Profile Update

<table>
<thead>
<tr>
<th>Report Period</th>
<th>Total Cases Reported*</th>
<th>Percent</th>
<th>Race Distribution (%)</th>
<th>Risk Group Distribution (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>&lt;13yrs</td>
<td>&gt;=13yrs</td>
<td>Total</td>
<td>Female</td>
</tr>
<tr>
<td>Latest 12 Months:</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>05/05-04/06</td>
<td>19.6</td>
<td>45.0</td>
<td>15.6</td>
<td>4.9</td>
</tr>
<tr>
<td>Five Years Ago:</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>05/06-04/02</td>
<td>25.3</td>
<td>37.4</td>
<td>8.5</td>
<td>3.0</td>
</tr>
<tr>
<td>Cumulative:</td>
<td>727</td>
<td>29,774</td>
<td>30,001</td>
<td>19.6</td>
</tr>
</tbody>
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