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Georgia Epidemiology Report

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

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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

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

Source: Adapted from Belay E., Schonberger L. Variant Creutzfeldt-Jakob Disease and Bovine Spongiform Encephalopathy. Clin Lab Med 2002;22:849-62.

*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 many have when investigating or reporting suspect/confirmed cases of CJD.

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

References

1 CDC: Index page: Prion Diseases. Retrieved February 08, 2006 from the Centers for Disease Control and Prevention website: http://www.cdc.gov/ncidod/dvrd/prions/index.htm

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/

3 Toward a Better Understanding of Human Prion Diseases. PLoS Med 3(2). Science Daily (via Veterinary Science Today) Retrieved February 07, 2006 from: http://www.vetscite.org/publish/items/002687/index.html

4 CDC: Fact Sheet: Variant Creutzfeldt-Jakob Disease. Retrieved March 2, 2006 from the Centers for Disease Control and Prevention website: http://www.cdc.gov/ncidod/dvrd/vcjd/factsheet_nvcjd.html

5 National Prion Disease Pathology Surveillance Center: About Human Prion Diseases/Acquired. Retrieved February 06, 2006 from the National Prion Disease Pathology Surveillance Center website: http://www.cjdsurveillance.com/abouthpd-acquired.html

6 National Prion Disease Pathology Surveillance Center: Welcome/Purposes of the Center. Retrieved February 06, 2006 from the National Prion Disease Pathology Surveillance Center website: http://www.cjdsurveillance.com/

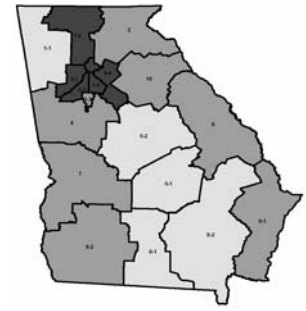
2005 Georgia Data Summary:
BREAST CANCER

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.

Breast Cancer Incidence Rates, Women, by Health District, Georgia, 1999-2002



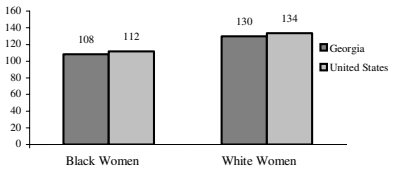
Rate significantly higher than the state

No significant difference from the state

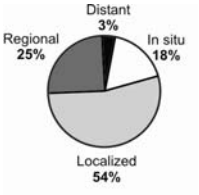
Rate significantly lower than the state

- The Northwest (1-1), South Central (5-1), North Central (5-2), South (8-1), and Southeast (9-2) Health Districts have significantly lower breast cancer rates than the state average.
- The North Georgia (1-2), Cobb/Douglas (3-1), Fulton (3-2), East Metro (3-4) and DeKalb (3-5) Health Districts have significantly higher breast cancer rates than the state average.

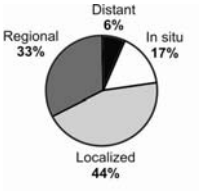
Age-adjusted Breast Cancer Incidence Rates, Georgia (1999-2002) and the United States (1998-2002)



Breast Cancer Incidence by Stage, Georgia (1999-2002)



WHITE WOMEN



BLACK WOMEN

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: DPH05.115H
Visit http://www.health.state.ga.us/programs/gccr/index.asp for more information about cancer in Georgia.

Georgia Department of Human Resources, Division of Public Health – 2 Peachtree Street, NW – Atlanta, GA 30303 – (404) 657-3103 – gdphinfo@dhr.state.ga.us – http://health.state.ga.us

The Georgia Epidemiology Report
Epidemiology Branch
Two Peachtree St., NW
Atlanta, GA 30303-3186

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Reported Cases of Selected Notifiable Diseases in Georgia Profile* for February 2006

Selected Notifiable Diseases	Total Reported for February 2006	Previous 3 Months Total Ending in February			Previous 12 Months Total Ending in February		
	2006	2004	2005	2006	2004	2005	2006
Campylobacteriosis	50	110	89	109	623	568	611
Chlamydia trachomatis	2076	7988	7969	7916	35169	34331	32966
Cryptosporidiosis	18	47	18	41	143	159	172
E. coli O157:H7	3	3	5	4	26	25	30
Giardiasis	25	176	174	127	828	896	719
Gonorrhea	879	3723	3777	3479	17161	16073	15376
Haemophilus influenzae (invasive)	10	34	42	30	92	126	100
Hepatitis A (acute)	4	113	40	13	740	266	110
Hepatitis B (acute)	10	119	92	29	597	447	155
Legionellosis	0	2	5	3	32	44	37
Lyme Disease	0	2	0	0	9	10	6
Meningococcal Disease (invasive)	1	11	8	3	32	17	13
Mumps	0	0	0	1	3	2	2
Pertussis	3	8	11	7	33	34	46
Rubella	0	1	0	0	1	0	0
Salmonellosis	55	279	239	250	2051	1925	2005
Shigellosis	60	164	102	169	1031	602	734
Syphilis - Primary	2	36	32	15	134	117	114
Syphilis - Secondary	15	124	120	63	480	477	465
Syphilis - Early Latent	11	126	76	53	695	366	352
Syphilis - Other**	32	177	225	126	860	895	794
Syphilis - Congenital	0	2	1	1	8	5	3
Tuberculosis	25	117	122	90	515	530	488

* 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

Report Period	Total Cases Reported*			Percent	Risk Group Distribution (%)						Race Distribution (%)		
	<13yrs	>=13yrs	Total	Female	MSM	IDU	MSM&IDU	HS	Blood	Unknown	White	Black	Other
Latest 12 Months:													
05/05-04/06	3	1,755	1,758	25.4	32.5	5.9	2.3	8.2	1.2	49.9	23.0	75.0	2.0
Five Years Ago:													
05/01-04/02	1	1,692	1,693	25.3	37.4	8.5	3.0	17.7	2.1	31.3	18.8	76.3	4.8
Cumulative:													
07/81-04/06	227	29,774	30,001	19.6	45.0	15.6	4.9	14.1	1.8	18.6	31.5	66.0	2.5

MSM - Men having sex with men **IDU - Injection drug users** **HS - Heterosexual**

* Case totals are accumulated by date of report to the Epidemiology Section