
Creutzfeldt-Jakob Disease Surveillance: Findings in New York City, 2001-2003

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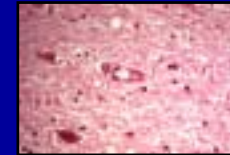
**New York City Department of
Health and Mental Hygiene**

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What is Creutzfeldt Jakob Disease (CJD) ?

- CJD is a Transmissible Spongiform Encephalopathy (TSE): a fatal degenerative disease of the central nervous system
- TSE's occur in humans and some animal species
- Primarily affecting older patients, mean age 62 years.
- Begins primarily with neurological symptoms- dementia, ataxia.

Human brain infected with CJD



Photomicrographs was prepared by Stephen J. DeAmond.

Characterized by microscopic vacuoles and amyloid plaques in gray matter of brain, causing brain to appear spongy.

What is variant CJD (vCJD)?

- A new form of TSE first diagnosed in the United Kingdom in 1995.
- Associated with eating contaminated meat products from cattle infected with Bovine Spongiform Encephalopathy (BSE) or “Mad Cow Disease”

- Begins primarily with psychiatric symptoms
- Primarily affecting young patients, mean age 28 years.
- Normal EEG

World Health Organization Criteria

Definite Case of CJD

- **Brain biopsy or autopsy**
 - ✓ **Western blot confirmed PrPsc**
 - ✓ **Immunocytochemistry**
 - ✓ **Presence of scrapie-associated fibrils.**

World Health Organization Criteria Probable Case of CJD

- ▶ Progressive dementia and at least two of the following:
 - Myoclonus
 - Visual or cerebellar signs
 - Pyramidal/extrapyramidal signs
 - Akinetic mutism
- ▶ Typical EEG during an illness of any duration; a positive result of a 14-3-3 CSF assay (measuring proteins associated with rapid loss of neuronal tissue) and a clinical duration to death of <2 years.
- ▶ Routine investigations should not suggest an alternative diagnosis.

World Health Organization Criteria Possible Case of CJD

- ▶ **Progressive dementia and at least two of the following:**
 - **Myoclonus**
 - **Visual or cerebellar signs**
 - **Pyramidal/extrapyramidal signs**
 - **Akinetic mutism**
- ▶ **No EEG or atypical EEG and a clinical duration to death of <2 years.**

TSE Surveillance in NYC

- In July 2001, TSEs were added to the reportable disease list in the New York City Health Code (Section 11.03) due to concerns about the potential introduction or emergence of vCJD in the United States.
- Based on national estimates, it was expected that the annual incidence of CJD would be approximately 1 per million population in New York City.

Surveillance Objectives

- **To review the current sources of CJD reports.**
- **To assess the incidence of CJD in New York City.**
- **To identify shortcomings in current CJD surveillance.**

Current Means of CJD Reporting

- Pathology reports from the National Prion Disease Pathology Surveillance Center at Case Western University in Cleveland, Ohio.
- Laboratory reports of elevated 14-3-3 proteins.
- Death certificates (listing CJD among the contributing factors)
- New York State Department of Health Alzheimer's Disease and Other Dementias' Registry.
- Provider reporting

National Prion Disease Pathology Surveillance Center

- **Established by Centers for Disease Control and Prevention (CDC) and the American Association of Neuropathologists.**
- **Offers free, state-of-the-art testing of CSF, blood and brain tissue to establish diagnosis of prion disease.**
- **Aids in arranging and paying for autopsies in suspect fatal cases.**

Methods of Case Review

- Review of medical records when available for New York City residents.
- Completion of CDC case abstraction form for all suspected patients < 55 years old and many selected patients > 55 years old.
- As applicable, assistance in arranging autopsies and transporting autopsy material to National Prion Disease Pathology Surveillance Center.

Reports of Suspected CJD in NYC

Since July 2001, 39 suspected cases of CJD have been reported to NYC DOHMH

<u>Source</u>	<u>N=</u>
National Prion Center	23
Laboratories	5
Death certificates	6
Alzheimers/Dementia Registry	4
Provider reporting	1

Reports of Suspected CJD in NYC

Of the 39 suspected cases reported:

NYC residents	22
Non-NYC residents*	17

* Non-NYC residents seeking care at NYC hospitals.

New York City Residents

N=22

Definite CJD cases: 6

Confirmed through pathologic examination of brain biopsy or autopsy tissue.

Probable and possible CJD cases: 10

CJD listed on death certificate/hospital chart despite lack of laboratory testing but met clinical criteria.

Ruled out as CJD: 5

Four with elevated 14-3-3 protein results but ruled out as CJD based on clinical criteria.

Pending 1

Incidence of CJD in NYC approximately

0.75/million. New CJD identified

New York City Residents

Definite and Probable/Possible Cases, N=16

Gender

Female 75%

Male 25%

Age

mean and median 71 years old

(range) 52-92 years

Race

White 75%

Unknown 19%

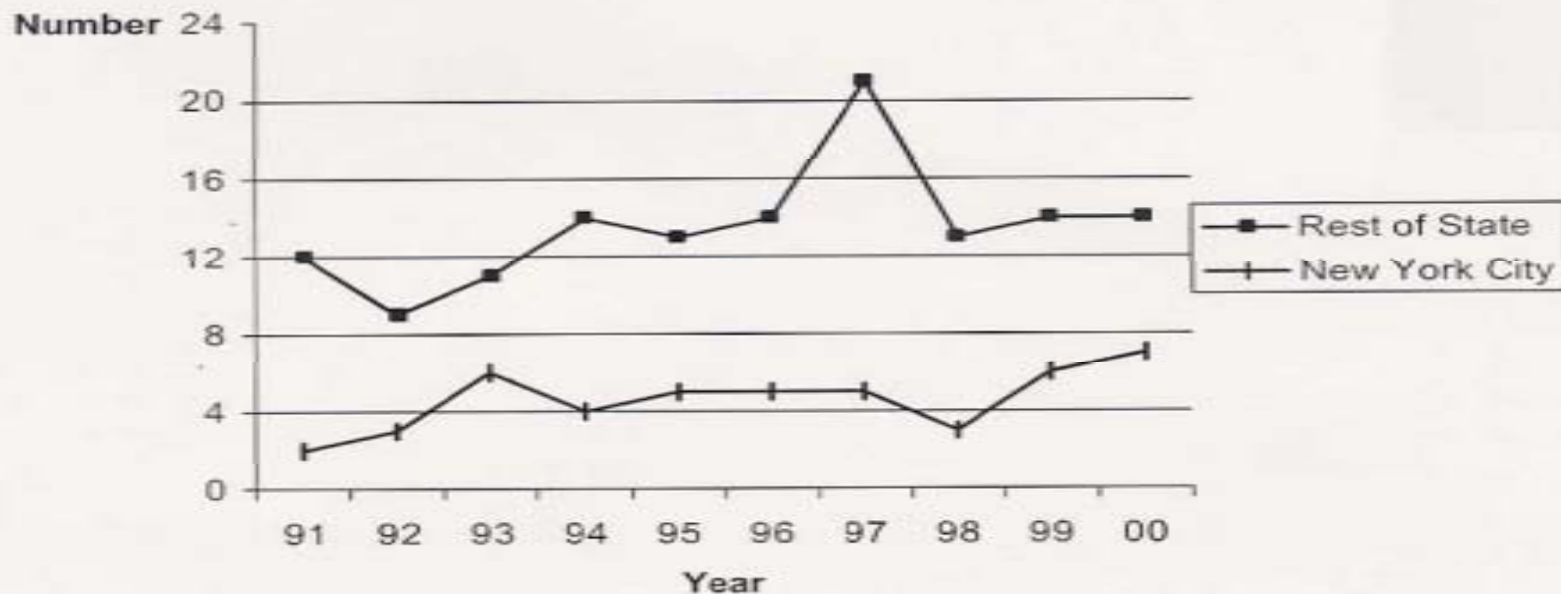
Black 6%

CJD in New York State (NYS) 1991-2000

- ▶ Average 18 CJD deaths/year.
- ▶ 17% of CJD deaths among persons <55 years old.
- ▶ NYC has 41% of NYS population but only 25% of CJD deaths.
 - ✓ From 1991-2000, NYC incidence of possible/probable/definite CJD was approximately .51/million.
 - ✓ 88% of cases were Caucasian.
 - ✓ Autopsies performed on approximately 25% of suspected cases.

CJD Deaths in New York State and New York City

Figure 1. Number of Deaths due to CJD Among New York State Residents, by Year and Region, 1991-2000



CONCLUSIONS

Sources of CJD Reports

- Most reports of probable/confirmed CJD are from the National Prion Disease Pathology Surveillance Center.
- Few reports from providers.

CONCLUSIONS

Incidence of CJD in NYC

- Surveillance efforts indicate that CJD incidence including possible/probable and definite cases (0.75/million) was lower than expected in NYC (1/million) in 2001-2003, but higher than incidence (0.51/million) seen in 1991-2000.
- Unclear if low incidence is the result of limited testing, low autopsy rate in general population, underreporting, or reflective of the true CJD incidence in NYC.

Recommendations

NYC DOHMH must intensify efforts to

- Educate providers about CJD/vCJD**
- Enhance provider reporting of suspected cases.**
- Encourage ante- and/or post-mortem testing**
- Educate providers on the free services offered by the National Prion Disease Pathology Surveillance Center**

Next Steps

- **A successful conference on CJD was held in Upstate New York in Oct. 2003. Plan for NYC conference to be held in late 2004.**
- **Educational update on TSE reporting and availability of free testing and autopsy sent to NYC providers and hospitals via our health alert system in Feb. 2004.**
- **Coordination with New York State and American Association of Neurologists to improve awareness of CJD/TSE.**