

Letter to the Editor:
**A Cluster of Creutzfeldt-Jacob Disease Patients
from Nassau County, New York, USA**

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Dear Editor

Creutzfeldt-Jacob disease (CJD) is a rare and transmissible neurological disorder, which has been increasing in frequency in the United States over the past two decades [1]. CJD is a spongiform encephalopathy characterized by a conformational change of prion protein [2]. The death rate for CJD in the USA is 0.9 per million [1].

We report 7 cases of CJD from North Shore University Hospital, a community-based teaching institution that serves Nassau County on Long Island, NY. These cases were diagnosed and died during the 12-mo period from mid-June 1999 to mid-June 2000. The estimated population of Nassau County in 1997 was 1,281,500, according to the New York State Department of Health [3]. These data suggest a CJD death rate in Nassau County of 5.5 per million, which is approximately 6 times the national rate. During the previous 1-yr period, no case of CJD was diagnosed in our laboratory.

The diagnosis of CJD in the 7 cases was based on pathological examinations performed by at least 2 pathologists. Three cases were confirmed by brain biopsy and 4 were diagnosed at autopsy. In 1 case,

CJD was not suspected during life. Pertinent data are summarized in Table 1. The patients' clinical histories were significant for rapidly deteriorating higher cortical functions, with confusion, speech disorders, nystagmus, ataxia, and/or myoclonus. Only 1 patient had a family history of CJD.

The autopsied brains were grossly unremarkable. At microscopic examination, all specimens showed the pathological features of CJD, including spongy degeneration, loss of neurons, and gliosis in the cerebral cortex. The basal ganglia were also involved. No Kuru plaques or inflammatory changes were noted. The patients' relatively advanced ages (median = 75 yr, range = 57 to 82 yr), the rapid course of their disease (median = 8 wk; range = 2 to 10 wk), and the absence of Kuru plaques suggest that these cases represent classical CJD, as opposed to new variant CJD [2].

The authors are concerned that the number of CJD cases in our catchment area appears to have increased dramatically during the 12-mo period. There is no clustering of the patients in any particular neighborhood. While the causes for the apparent increase of CJD cases are unknown, enhanced alertness for the disease and early recognition of its symptoms may be possible explanations. The cluster of CJD patients ought to be investigated thoroughly, which is beyond the scope of this letter.

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Table 1. Available information regarding the patients with Creutzfeldt-Jacob disease from Nassau County, New York.

Patient #	Age (yr)	Sex	Duration of symptoms (wk)	Family history of CJD	Signs & symptoms	Method of diagnosis	Other findings
1. (BP)	82	M	8	none	no response to commands, speech	autopsy	EEG, CT, LP negative
2. (DE)	75	M	2	none	ataxia, dysarthria, nystagmus	autopsy	na
3. (ER)	68	M	8	father	memory loss, jerking motion of extremities	autopsy	basal flare in MRI
4. (EL)	81	M	20	none	post-operative progressive confusion, dementia	biopsy	CT diffuse atrophy; no focal lesion
5. (LH)	57	F	10	none	progressive loss of cognition, myoclonus, ataxia	autopsy	MRI non-contributory, EEG ↑amplitude
6. (MW)	73	F	6	none	abnormal speech and movement	biopsy	na
7. (AS)	75	F	8	none	rt 3rd nerve palsy, progressive encephalopathy	biopsy	CT, MRI - meningioma

na = not available

References

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2. Wehl CC, Roos RP: Creutzfeldt-Jacob disease, new variant Crutzfeldt-Jacob disease, and bovine spongiform encephalopathy. *Neurol Clin* 1994;4:835-859.
3. New York State Department of Health. Internet Home Page, <http://www.health.state.ny.us/nysdoh/vs97/tab03.htm> (downloaded on 8 December 2000).