Idaho Disease

BULLETIN

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Reported Creutzfeldt-Jakob Disease (CJD) in Idaho

CJD was not reportable in Idaho until 2004. Prior to that, information on the number of cases was based entirely on death certificates. Among Idaho residents in the last 20 years, there have been 25 deaths reported on death certificates as caused by CJD (2004 data is preliminary). The annual number of reported CJD deaths ranged from zero to three. CJD deaths were reported from 17 Idaho counties. From 1985 to 2004, only 10 of 25 (40%) persons with CJD on their death certificate received an autopsy to confirm the clinical diagnosis. Nationally only about 30 percent of cases are autopsied.

CJD in Idaho: 2005

South Central Idaho
Five possible CJD cases reported between February and July 2005 have been under investigation by the South Central District Health Department and the Idaho Department of Health and Welfare, Office of Epidemiology and Food Protection (OEFP). All five patients are deceased.

Of the five people who died from possible CJD, autopsies were conducted on three, and brain samples were sent to the National Prion Disease Pathology Surveillance Center (NPDPSC) for testing. Results as of 10/11/2005 are:

- Preliminary results indicated a prion disease, likely CJD in one person.
- Final results indicated that one person did not have a prion disease and one person had sporadic CJD.

Investigation continues on the two patients who have not been autopsied. This includes a review of medical records and interviews with treating physicians. A survey on residence, travel, dietary habits, occupation, surgeries, and other experiences was conducted with family members.

South Central Idaho Investigation Findings
Of the four possible CJD cases still under investigation, all patients were white females, over 55 years of age (mean 70.8 years). No common ethnic population of origin could be identified. The estimated time from onset of illness to death ranged from 1.4 to 10.6 months (mean 4.3 months); however, the exact date of onset of illness could not be established in some cases.

All patients lived in Twin Falls or Minidoka County in the South Central public health district at the time of death; one patient also lived in a third Idaho county earlier in life. The number of years of continuous residence in Idaho prior to death ranged from less than one year to 72 years (mean 49 years). One patient lived for 18 months in Great Britain prior to 1980. No other travel to Europe was reported for any patient, and no travel in common to all four patients was found.

No reported surgical procedures were common to all four patients, and no patients had neurologic surgery or transplants. The number of reported surgeries over a patient's lifetime ranged from 1 to 3 (mean 2). No patients had been injected with growth hormone.

Additional reports of possible CJD
A possible case of CJD in an Elmore County male over 55 was reported to the Central District Health Department in August. This patient is deceased, and brain tissue tested negative for prion disease by the NPDPSC. Two reports from southeastern Idaho about deceased patients over age 55, one female and one male, neither of whom was autopsied, are being investigated by the Southeastern District Health Department. A report from northern Idaho about a female patient under age 55, who is deceased, was being investigated and initial results from NPDPSC are consistent with prion disease.

For further information and updates, see the Idaho Department of Health and Welfare's CJD web page at: http://www.diseaseinfo.idaho.gov .

Diagnostic Services for Prion Disease Evaluation

Currently prion diseases can be definitively diagnosed only by examination of brain tissue, usually obtained at autopsy. Barriers to autopsy include lack of patient and family education about the necessity for autopsy to obtain a definitive diagnosis, religious beliefs and personal wishes of the patient or patient's family, cost considerations, and difficulty finding pathologists willing to autopsy a suspect case of transmissible spongiform encephalopathy (TSE). The OEFP and local public health districts are collaborating with the NPDPSC to help remove some of the barriers to autopsy of suspected TSE cases.

The NPDPSC was established by the Centers for Disease Control and Prevention and the American Association of Neuropathologists in 1997. It is the national reference center for prion diseases, providing advanced neuropathologic and biochemical diagnostics including histopathology, immunohisto-chemistry, Western blot, and prion gene analysis, which is used to distinguish the type of prion disease (familial, sporadic, or acquired). To augment antemortem prion disease diagnostics, CSF can be submitted to determine the presence of the protein marker 14-3-3, though this is not a confirmatory test.

All testing at NPDPSC is free of charge. Results are reported to the health care provider and, as required by state law, to the OEFP. In addition, NPDPSC can help make arrangements for a brain-only autopsy, including providing a pathologist if none is available locally. All expenses including transporting of the body to the reference institution, collecting brain tissue, returning the body, shipping tissues to the NPDPSC, and testing brain tissue at NPDPSC will be covered, when necessary.

Improving Prion Disease Surveillance

Health care providers can work to improve prion disease surveillance in Idaho by following these steps:

1. Report all suspected cases of human prion disease to the OEFP (208-334-5939) or your local public health district within three days of when the diagnosis is suspected or confirmed, and participate in a confidential case investigation to document demographic and clinical information and help determine possible sources or risk factors for illness.
2. Discuss autopsy with the patient's family. With their permission, post-mortem arrangements can be made through the NPDPSC for a brain-only autopsy at a referral institution.
3. Call the NPDPSC (216-368-0587) if you would like to use their laboratory or autopsy services. Specimen collection and shipping instructions can be found at http://www.cjdsurveillance.com.
4. Please indicate on the patient's death certificate the diagnosis of CJD or other TSE when applicable and whether or not an autopsy was performed. If awaiting autopsy results, the entry in the *Cause of Death* section may be entered as "Pending autopsy results"; the *Manner of Death* must be completed as "Natural". Upon completion of autopsy, a Supplemental Information for Cause of Death form, provided by the Idaho Department of Health and Welfare Bureau of Health

Policy and Vital Statistics, should be completed and submitted by the physician who signed the death certificate.

Resources for Families of CJD Patients

To support patient families, the CJD Foundation operates a national toll-free line (800-659-1991) and a web site: http://www.cjdfoundation.org/. Other family support resources can be found at http://www.rarediseases.org/search/rdbdetail_abstract.html?disname=Creutzfeldt+Jakob+Disease.

CJD Reference Material

Centers for Disease Control and Prevention: http://www.cdc.gov/ncidod/dvrd/cjd/

The National Creutzfeldt-Jakob Disease Surveillance Unit (UK): http://www.cjd.ed.ac.uk/index.htm

Medscape article on prion diseases (must register to view): www.medscape.com/viewarticle/410863

Influenza: A New Strategy for Prioritizing Vaccinations

The vaccine supply for the 2005-06 influenza season is still up in the air because of uncertainties regarding production of vaccine, exact number of available doses, and timing of vaccine distribution. Four manufacturers expect to provide vaccine to the U.S. market. Sanofi Pasteur, Inc., projects production of up to 60 million doses of inactivated influenza vaccine; Chiron Corporation 18–26 million doses; GlaxoSmithKline, Inc., eight million doses. MedImmune Vaccines, Inc., producer of the nasal-spray influenza vaccine (also called live attenuated influenza vaccine, or LAIV), projects having approximately three million doses available for distribution.

To ensure that people who are at highest risk of complications from influenza have access to vaccine, CDC recommends certain priority groups receive inactivated influenza vaccine until Oct. 24, 2005. Beginning Oct. 24, 2005 all persons were eligible for vaccination.

The following are the priority groups which should have been targeted to receive inactivated influenza vaccine prior to Oct. 24, 2005:

- Persons aged 65 years and older, with and without chronic health conditions;
- Residents of long-term care facilities;
- Persons aged 2–64 years with chronic health conditions;
- Children aged 6–23 months;
- Pregnant women;
- Health care personnel who provide direct patient care;
- Household contacts and out-of-home caregivers of children under 6 months; and
- Evacuees from hurricane Katrina.

It should be noted that vaccination with the live, nasal-spray flu vaccine (FluMist®) is always an option for healthy people 5–49 years who are not pregnant. This vaccine is not subject to prioritization and can be given to healthy individuals 5–49 years at any time.

Demand for influenza vaccine typically falls off quickly after November, even when there is a shortage. The current approach was developed to balance two competing priorities:
(1) Ensuring an ample opportunity to vaccinate people at highest risk of complications from influenza, providers who care for them, and close contacts of children under 6 months of age; and
(2) Allowing ample time to vaccinate other priority groups and those desiring vaccination before demand declines. Community vaccinators and health officials generally need at least four to five weeks for optimal planning efforts. Oct. 24, 2005, was selected as the best date to achieve a balance between these priorities, and, therefore, the priority group restrictions were lifted on this date.

Thank You to Hurricane Katrina Volunteers

Public health staff at the IDHW and the local public health districts would like to thank the many physician and other health care provider

volunteers who signed up to assist with evaluating and treating evacuees from Hurricane Katrina. While the crowds did not materialize in Idaho as anticipated, individuals and families have arrived through faith-based organizations, family contacts, permanent relocation, and other means. Some ongoing activities have occurred to assure these displaced people receive health care. We are very grateful to the private provider community for stepping forward to assist when requested on short notice. It is our hope that you will remain available in the future if surge capacity is required in response to another public health crisis.

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