1. Surveillance for Human Transmissible Spongiform Encephalopathy

Transmissible Spongiform Encephalopathies (TSEs) are a family of fatal neurodegenerative diseases affecting both humans and animals. TSEs are caused by an abnormally configured form of a naturally-occurring cellular protein called a prion. Human TSEs include sporadic Creutzfeldt-Jakob Disease (sCJD), variant CJD, and several inheritable or familial forms. Reports of TSEs are investigated by the Wisconsin Division of Public Health (DPH) rather than local health departments. This article summarizes TSE surveillance in Wisconsin for the interval 2003-2012. Note that the term “TSE” and “human prion diseases” are used interchangeably in this summary.

Background:
During 2002, chronic wasting disease (CWD) was discovered among free-ranging whitetail deer in Wisconsin. This finding occurred soon after the outbreak of human variant CJD in Europe during the 1990s and the detection of North American cattle with bovine spongiform encephalopathy. All these factors provided the impetus for the DPH to institute a surveillance system for human prion diseases. Surveillance began during 2003 when DPH received funding under the ELC Cooperative Agreement with the Centers for Disease Control and Prevention (CDC) to provide support for surveillance of human TSEs. Human prion diseases became officially notifiable in Wisconsin in March, 2008. All case investigations are performed by DPH staff; LHDs are not responsible for TSE follow up.

The primary goal of this surveillance is to identify and accurately classify all cases of human prion disease that occur in Wisconsin. Case finding is accomplished by 1) ongoing review of death certificates, 2) investigation of reports from health care providers, and 3) investigation of potential cases identified by tests of cerebrospinal fluid for the tau and 14.3.3 proteins conducted at the National Prion Disease Pathology Surveillance Center (NPDPSC) in Ohio. These protein markers, although not pathognomonic, are strongly suggestive of CJD in a patient with compatible clinical features.

When a suspected case is identified, case confirmation and classification are conducted by reviewing patient medical charts and facilitating brain-only autopsies whenever possible. Case confirmation requires histologic examination of brain tissue and Western blotting of any abnormal prion protein detected. Post-mortem examinations are encouraged through conversations with clinicians, social workers and family members. The CDC provides funding for autopsy services, the transport of remains and all testing services. Autopsy specimens are forwarded to the NPDPSC. The current World Health Organization and CDC case definitions for confirmed, probable and possible cases are applied. (See www.cdc.gov/ncidod/dvrd/cjd/resources/CDCs_Diagnostic_Criteria_for_CJD-2010.pdf).

Surveillance Results:
94 cases of CJD were reported among patients who died between 2003 through 2012 (Figure 1). Of these cases, 64 were confirmed (62 sporadic CJD and 2 familial CJD), 25 were probable and 5 were possible. The 64 confirmed cases represent an average annual incidence of approximately 1.1 case per million population. Confirmed cases occurred among residents of 27 counties (Figure 2).
Among the 62 patients with cases of sCJD, 33 (53%) occurred in females, mean age was 67.5 years (range 43-88 years), and the mean duration of illness was 7 months (range 3 weeks-33 months). No pathology results suggestive of variant CJD were received.

Figure 1

Summary of Wisconsin CJD Surveillance, 2003 - 2012

(N = 94) 64 confirmed: 62 sporadic CJD, 2 familial CJD

Figure 2

Confirmed CJD cases by county of residence, Wisconsin, 2003–2012 (n=64)
Registry of Persons Who Consumed Venison from CWD-positive Deer:

Although the prion that causes chronic wasting disease (CWD) has not been shown to be pathogenic for humans, the possibility that cross species transmission to humans may occur cannot be absolutely ruled out. Accordingly, the DPH advises the public not to consume venison from deer which test positive for CWD. Despite this recommendation, some people do eat venison from known CWD-positive deer. With the assistance of the DNR, the DPH attempts to contact these individuals and obtain identifying information. This process has been ongoing since the 2003 hunting season.

Currently, there are 1,017 venison consumers included in this registry for whom we have at least a first and last name and the town of residence. When new CJD cases are reported to DPH, the patient name is cross-checked against this registry. To date, no CJD patients have matched with any of the registry names. Because of the extraordinarily long incubation period of prion diseases, this project will be ongoing well into the future if funding permits.

Questions about prion disease surveillance or suspect case reports can be directed to Jeannie Druckenmiller (608/516-5847) or Jim Kazmierczak (608/266-2154) in the Communicable Disease Epidemiology Section. Although case follow up is conducted by DPH, LHD staff and providers are asked to report any suspect cases promptly so autopsy consent and other logistics can be arranged prior to the patient’s death.

2. RABIES JURISDICTIONAL ISSUES – WHOSE JOB IS IT ANYWAY?

The Communicable Disease Epidemiology Section frequently receives questions about roles and responsibilities when multiple local health departments (LHDs) are involved in an animal bite incident follow up. The typical scenario involves a bite that occurs in County A, but the animal and owner reside in County B. Assuming a quarantine is needed, which agency issues the quarantine order? Who ensures compliance? And who serves as the point of contact for the patient and the medical provider?

The general rule is: **Issuing an animal quarantine and ensuring compliance with the quarantine is the responsibility of the jurisdiction in which the animal resides.**

- In the scenario above, an official, provider, or citizen from County A should report the bite to the health department or law enforcement agency in County B where the animal resides. At that point the County B official issues the quarantine order as per s.95.21(4)(a) which states “an officer shall order a dog or cat quarantined if the officer has reason to believe that the animal bit a person, is infected with rabies or has been in contact with a rabid animal.”

- County B officials should then notify the LHD in County A (i.e., the county in whose jurisdiction the bite victim resides) that the offending animal has been quarantined, because this should eliminate any need to consider rabies post-exposure prophylaxis for the bite victim.

- Upon the animal’s release from the quarantine, County B officials should notify the LHD in County A (where the bite victim resides) that the quarantine has been satisfactorily completed. The County A LHD can then notify the bite victim and the provider.

- If a Wisconsin resident is bitten by an animal that resides in another state, contact the Communicable Disease Epidemiology Section. Section staff can work with other state health departments to ensure the animal is appropriately quarantined and observed.
3. LOCAL HEALTH DEPARTMENTS COLLABORATE WITH STATE’S SURVEILLANCE AND OUTBREAK SUPPORT TEAM TO ENHANCE ENTERIC DISEASE SURVEILLANCE

During fall 2009, the Wisconsin Division of Public Health established the Surveillance and Outbreak Support (SOS) Team to improve timely routine surveillance for *Salmonella*, Shiga-toxin producing *E. coli*, and *Listeria* (SSL) and expand the state and local health departments’ (LHDs) capacity during outbreak investigations. The SOS team is comprised of graduate students in the UW School of Medicine and Public Health who work under the direction of a DPH epidemiologist.

Through sustained efforts at the local level and increased interviewing capacity provided by the SOS Team, an increasing percentage of cases of SSL infection had completed interviews with food histories each year. Among cases of SSL infections reported during 2010-2012, the percentage with a completed routine interview with food history increased from 73% of 1080 cases during 2010, to 79% of 1,081 SSL cases during 2011, and 83% of 1,147 cases during 2012 (see Figure). As a result, when illness clusters are detected, exposure information is often already available for review by epidemiologists. Currently, the median time elapsed from receipt of a case report to a completed interview is 2 days for *Salmonella* and STEC infections, and 3.5 days for listeriosis.

In addition to the interviews completed for SSL surveillance, the SOS Team provides interviewing support for outbreak-related and other enteric disease surveillance. The SOS Team completed 327 interviews during 2010, 497 during 2011, and 622 during 2012.

LHDs that need assistance in conducting enteric disease follow-up resulting from emergent situations should contact Justin Kohl in the Communicable Disease Epidemiology Section (608/ 266-2701 or justin.kohl@wi.gov).

Completed SSL interviews by agency

![Completed SSL interviews by agency](image)
4. RESOURCES FOR MENINGOCOCCAL DISEASE AND INVASIVE HAEMOPHILUS INFECTIONS

We want to remind healthcare practitioners that the occurrence of invasive *Neisseria meningitidis* infection (meningococcal disease) and invasive *Haemophilus influenzae* infection (all serogroups, including type b) typically increases during the cold weather months. *Neisseria meningitidis* and *H. influenzae* can both cause sepsis or meningitis resulting in serious illness and death. Both diseases are DPH **Category 1** diseases because of the severity of illness and the need to identify and treat direct contacts immediately. Please forward all invasive *Haemophilus influenzae* and *Neisseria meningitidis* isolates to the Wisconsin State Laboratory of Hygiene for serotyping. Shipping and testing are conducted at no cost to the submitting laboratories.

The uptake of highly efficacious conjugate vaccines for the prevention of meningococcal disease and invasive *H. influenzae* type b (Hib) infections has substantially reduced the annual reported number of cases among Wisconsin residents. However, even a single case can be challenging to investigate. To assist with such investigations, the Communicable Disease Epidemiology Section has developed protocols and flowcharts for both diseases. This guidance should help an investigator identify reportable cases, and provide instructions for isolate submission and contact identification and prophylaxis. These resources also provide DPH contacts for assistance during the investigation.

Protocols and flowcharts can be printed and shared with any health care practitioner who may benefit from this information. Please refer to the following links on the Department of Health Services website for the protocols and flowcharts.

*Haemophilus influenzae:*

*Neisseria meningitidis:*
http://www.dhs.wisconsin.gov/communicable/InvasiveBacteria/PDFfiles/MeningFlowchart.pdf

5. **HEALTHIEST WISCONSIN 2020 BASELINE AND HEALTH DISPARITIES REPORT** RELEASED

The Wisconsin Department of Health Services has released the *Healthiest Wisconsin 2020 Baseline and Health Disparities Report*. This report provides baseline data about populations experiencing health disparities in Wisconsin and includes factors such as race/ethnicity, socioeconomic status, level of urbanization, sexual preference, and gender identity. Users can orient themselves to this large report by reviewing the text on the web page and the Executive Summary.

The report is available at [www.dhs.wisconsin.gov/hw2020/hw2020baselinereport.htm](http://www.dhs.wisconsin.gov/hw2020/hw2020baselinereport.htm). Please provide your contact information under the “Help us evaluate the report” link in the Spotlight box so DHS staff can follow up with you later to solicit your feedback regarding the report.

Please forward this announcement to others interested in public health in Wisconsin.
6. UPCOMING MEETINGS, TRAININGS & IMPORTANT DATES

- **Tuberculosis Summits and Trainings**
  - April 11, Minocqua WI: TB 101
  - April 24, Verona WI: Southern Region TB Summit
  - April 28, Milwaukee WI: Southeast Region TB Summit
  - April 30, Eau Claire WI: Western Region TB Summit
  Contact Lorna Will, BCDER (608-261-6319 lorna.will@wi.gov)

- **Communicable Diseases Spring Seminars**
  - June 10, Oconomowoc
  - June 11, Minocqua
  - June 12, Eau Claire
  Contact Karen Kopetskie, BCDER (608-264-6060 karen.kopetskie@wi.gov)

- **Wisconsin APIC State Conference, May 1-2, Country Inn and Waterpark, Pewaukee WI**
  Contact Gayle Land, Wheaton Franciscan Healthcare (414-447-3511 gayle.land@wfhc.org)

- **Vectorborne Disease Surveillance Seminar, April 10, Merrill, WI**
  Presentations targeted towards: Public Health Staff (9:00-11:00), Medical Providers (12:00-1:00), General Public (4:00-5:30)
  Contact: Kristi Krombholz, Lincoln County HD (715-539-1376 kkrombholz@co.lincoln.wi.us)

The Wisconsin Epi Express is posted online at [http://www.dhs.wisconsin.gov/communicable/WiEpiExpress/Index.htm](http://www.dhs.wisconsin.gov/communicable/WiEpiExpress/Index.htm) and distributed by email to local, tribal, regional and state public health officials and infection preventionists in Wisconsin. Suggestions for article topics are welcomed. Distribution list removal or addition requests and topic suggestions should be sent to: Barb Anderson: Barb.Anderson@wi.gov.