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## **A National Assessment of Amyotrophic Lateral Sclerosis And Other Chronic Neurologic Conditions State Surveillance Programs**

A Report for the  
Agency for Toxic Substances and Disease Registry (ATSDR)

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## **Executive Summary**

On October 8, 2008, the President signed the ALS Registry Act (Public Law No. 110-373) into law. It authorizes the Centers for Disease Control and Prevention (CDC) to establish a national amyotrophic lateral sclerosis (ALS) patient registry to identify ALS cases in the United States. The Agency for Toxic Substances and Disease Registry (ATSDR) has already begun to create the building blocks for this national registry by finding partners to pilot projects that would provide information for developing strategies to identify patients and highlight best practices in data sharing and acquisition.

In November, ATSDR requested partnership with CSTE in assessing all existing state chronic neurologic disease surveillance systems, specifically concerning ALS. In collaboration with ATSDR and several CSTE member epidemiologists, CSTE created an assessment to be sent to the states.

Nine (18%) states reported currently conducting surveillance for neurologic diseases. Only South Carolina conducted surveillance for all of the chronic neurologic diseases. Two states each reported conducting surveillance for only one neurologic disease: Alzheimer's disease (South Dakota) and ALS (Massachusetts).

This report compiles results of the assessment conducted by CSTE about the surveillance of chronic neurologic diseases, including ALS, by state. All information was provided by state health department staff specializing in chronic disease or environmental health epidemiology. The results should prove useful in determining the geographic distribution of surveillance activities and the legal impetus for surveillance; identifying the type of noninfectious, nontraumatic neurologic disease surveillance conducted by states; and providing further information about state programs associated with ALS surveillance.

## **Introduction**

The Council of State and Territorial Epidemiologists (CSTE) promotes the effective use of epidemiologic data to guide public health practice and improve health. CSTE accomplishes this by supporting the use of effective public health surveillance and good epidemiologic practice through training, capacity development, and peer consultation; developing standards for practice; and advocating science-based policy.

Senate Bill 1353 amends the Public Health Service Act to provide for the establishment of an amyotrophic lateral sclerosis (ALS) patient registry. The ALS Registry Act amends the Public Health Service Act to require the Director of the Centers for Disease Control and Prevention (CDC) to 1) develop a system to collect data on ALS and 2) establish a national registry for the collection and storage of ALS data.

On October 8, 2008, the President signed the ALS Registry Act (Public Law No. 110-373) into law. It authorizes the Centers for Disease Control and Prevention (CDC) to establish a national amyotrophic lateral sclerosis (ALS) patient registry to identify ALS cases in the United States.

The aim of a national ALS registry is to use the data collected to help discover the cause of and develop treatments for this fatal neurologic disease. CSTE supports meaningful scientific inquiry and surveillance for chronic neurologic diseases while recognizing that strong national data must be collected through partnerships between state and local health departments. Dissemination of these public health data will arm scientists with the tools necessary to search for a means to mitigate the effects of ALS and to move forward in finding a cure.

In November 2008, the Agency for Toxic Substances and Disease Registry (ATSDR) requested partnership with CSTE in identifying and assessing all existing state neurologic disease surveillance systems, particularly those concerning ALS. This report explains the method of assessment and presents results for the nine states that currently conduct surveillance for chronic neurologic diseases. It also includes information obtained from interviews with epidemiologists in the two states that maintain an ALS disease-specific surveillance system. ATSDR can use the information in this report to help meet the objectives of the ALS Registry Act.

## **Method of Assessment**

### *Web-Based Assessment*

An assessment tool was developed to examine the chronic neurologic disease surveillance activities of states using Survey Monkey (SurveyMonkey.com; 2009). It was administered to the CSTE identified state health department points of contact in chronic disease and environmental health. The assessment consisted of seven questions (Appendix A). Chronic Disease and Environmental Health Epidemiology points of contact that CSTE maintains for each state health department were contacted and

instructed to provide one response per state. In the three instances when both points of contact responded to the assessment, follow-up was conducted and discrepancies resolved.

From late February through mid-March 2009, CSTE conducted the ALS State Surveillance Assessment. The assessment asked respondents to indicate whether their state conducted surveillance for chronic neurologic diseases in general and for ALS in particular. States without ALS surveillance systems were asked via the assessment whether they were considering adding surveillance for any chronic neurologic diseases. For states conducting surveillance activities for chronic neurologic diseases, data were collected on specific neurologic diseases and conditions: motor neuron disease (e.g., ALS), multiple sclerosis, Parkinson's disease, Alzheimer's disease, epilepsy, Guillain-Barré syndrome, Creutzfeldt-Jakob disease, and other. If a state indicated conducting surveillance for a neurologic disease, follow-up questions were asked to determine whether the disease was a reportable condition and whether there was a state legislative mandate to conduct the surveillance.

#### *Follow-Up Telephone Interview*

ATSDR and CSTE collaborated on the questions for a follow-up interview of states that reported conducting ALS surveillance (Appendix B). CSTE epidemiology staff conducted the telephone interviews with the individuals who maintain the surveillance systems. The goal of the telephone interviews was to obtain general information about state surveillance programs and associated activities.

### **Results**

The majority of respondents to the assessment were Chronic Disease and Environmental Health Epidemiology points of contact in each state. Their positions/titles were as follows: chronic disease epidemiologist, branch chief, state epidemiologist, state toxicologist, director of surveillance, chief medical officer, and other. The program areas in which respondents are employed within the Department of Health also varied. In most cases, respondents were the points of contact for the surveillance system.

Of the neurologic diseases for which states conduct surveillance, Creutzfeldt-Jakob disease was conducted most often and is the neurologic disease with the most state legislative mandates for its surveillance. Four states reported collecting Alzheimer's disease surveillance information, and four collect Parkinson's disease surveillance information. Three states reported collecting surveillance data for both Alzheimer's disease and Creutzfeldt-Jakob disease.

Surveillance for chronic neurologic disease varied from state to state (Table 1). Both Delaware and Idaho reported collecting information on Guillain-Barré syndrome and Creutzfeldt-Jakob disease. Only South Carolina conducted surveillance for all of the

chronic neurologic diseases. Two states each reported conducting surveillance for only one neurologic disease: Alzheimer's disease (South Dakota) and ALS (Massachusetts).

**Table 1. Surveillance for chronic neurologic diseases, by disease and state - ALS State Surveillance Assessment, 2009**

State	Motor neuron disease (e.g., ALS)	Multiple sclerosis	Parkinson's disease	Alzheimer's disease	Epilepsy	Guillain-Barré syndrome	Creutzfeldt-Jakob disease
California	no	no	<b>yes</b>	<b>yes</b>	<b>yes</b>	no	<b>yes</b>
Delaware	no	no	no	no	no	<b>yes</b>	<b>yes</b>
Idaho	no	no	no	no	no	<b>yes</b>	<b>yes</b>
Massachusetts	<b>yes</b>	no	no	no	no	no	no
Nebraska	no	no	<b>yes</b>	no	no	no	<b>yes</b>
New Mexico	no	<b>yes</b>	<b>yes</b>	<b>yes</b>	no	no	no
New York	no	no	no	<b>yes</b>	no	no	<b>yes</b>
South Carolina	<b>yes</b>	<b>yes</b>	<b>yes</b>	<b>yes</b>	<b>yes</b>	<b>yes</b>	<b>yes</b>
South Dakota	no	no	no	<b>yes</b>	no	no	No

For each of these chronic neurologic diseases, states varied with respect to legislative mandates, the reportability of the diseases, and implementation of surveillance systems (Table 2). For example, five states conduct surveillance for Alzheimer's disease and only one state listed Alzheimer's disease as reportable and as being mandated.

Several states conduct disease surveillance without considering the condition reportable or having a legislative mandate to conduct surveillance. Only Massachusetts reported a condition as reportable but does not presently conduct surveillance on that condition (Multiple Sclerosis).

**Table 2. States conducting surveillance for chronic neurologic disease (n = 9) - ALS State Surveillance Assessment, 2009**

Disease	No. states conducting surveillance	No. states for which condition is reportable	No. states for which surveillance is legislatively mandated
Alzheimer's disease	5	1	1
Creutzfeldt-Jakob disease	6	4	4
Epilepsy	2	0	0
Guillain-Barré syndrome	3	1	1
Motor neuron disease (e.g., ALS)	2	1	1
Multiple sclerosis	2	1*	0
Parkinson's disease	4	2	2

\*In Massachusetts, multiple sclerosis is a reportable condition, but surveillance activity for it is not conducted.

## State Interviews

### *Massachusetts*

Christine Fischetti, Epidemiologist/Registry Coordinator  
Massachusetts Department of Public Health

Massachusetts began analyzing ALS surveillance data in January 2008. The state used passive surveillance to collect data from 2007, with the intention of annually reviewing the previous year's data. Massachusetts conducts passive surveillance by requesting all (approximately ~600) neurologists in the state to report diagnoses of ALS, muscular atrophy, and bulbar palsy. Furthermore, Massachusetts collects cases from nine major hospitals and from death certificates, hospices, and ALS advocacy group lists. Massachusetts has a legislative mandate to conduct ALS surveillance and uses *International Classification of Diseases, Ninth Revision* (ICD-9) codes for ALS, muscular atrophy, and bulbar palsy as case definitions. The state uses ICD-10 codes to find cases from death certificates.

Massachusetts developed its surveillance system by using information from five studies. The first study determined the prevalence of ALS in southeastern Massachusetts. A second study examined the feasibility of various options for a statewide ALS surveillance system. The third and fourth studies were pilot studies of an ALS registry conducted first in Essex County and then in Boston. The last study examined feedback from focus groups about implementing an ALS surveillance system in Massachusetts. These studies are detailed in the Massachusetts protocol.

Massachusetts learned many lessons from the above studies. One major lesson was the need to identify a method to capture only true ALS cases; unlike for cancer, no central source exists for identifying ALS patients. In addition, ALS often is difficult to diagnose. To address this problem, the Massachusetts registry required a two-stage data abstraction process, as follows: 1) nurses with neurologic training established the eligibility of a case (based on patient residence, date of diagnosis, and certainty of diagnosis) according to the El Escorial diagnostic criteria; and 2) additional clinical information was collected and the diagnosis was confirmed by an independent neurologist according to the criteria of the *World Federation of Neurology*.

Learning the feasibility of establishing such a registry was valuable. The average length of time for medical record abstraction was 1–3 hours per record, excluding travel time. It is often required that surveillance personnel obtain multiple records from different institutions. Finally, the patients themselves may play an important role in ensuring the completeness of any registry; if ALS advocacy groups publicize the existence of an ALS registry, patients can remind their physicians to submit information.

In addition, Massachusetts had to address legal hurdles. Massachusetts amended some state regulations to encourage reporting of cases and ensure protection of privacy and compliance with the Health Insurance Portability and Accountability Act. An advisory committee was recommended to support the operations of the registry once

implemented; consisting of neurologists from ALS centers, representatives from ALS advocacy groups, patients and their caregivers, and providers. ALS surveillance remains ongoing within Massachusetts. For additional information, see the attached report, *Massachusetts ALS Surveillance* from the state's Department of Public Health.

*South Carolina*

Khosrow Heidari, Chronic Disease Epidemiologist  
Department of Health and Environmental Control, South Carolina

South Carolina began its ALS surveillance system in 2001 because of increased complaints regarding the occurrence of chronic neurologic disease within local communities. Surveillance covers the entire state. Records are acquired from all South Carolina hospitals, emergency departments, and ambulatory surgery centers. No legislative mandate exists in the state, and ALS is not reportable. South Carolina uses ICD-9 and ICD-10 codes to passively collect ALS data.

Annually, all hospital discharge, emergency department, and ambulatory surgery center records are sent to the Office of Research and Statistics at the State Budget and Control Board, where information is de-identified. Then the Office of Chronic Diseases Epidemiology and Evaluation scans the information for evidence of ALS diagnosis. Other variables collected by the surveillance system include duration of hospital stay, charges, comorbidities, specialty care and services received, source of payment, basic demographic data, and county of residence. No standard or protocol for ALS surveillance exists in South Carolina. Surveillance for chronic neurologic disease remains ongoing.

## Discussion

Because the initial inquiry about where ALS surveillance activities fall within a given state health department yielded no single answer, CSTE sent the ALS State Surveillance Assessment both to chronic disease and environmental health epidemiology points of contact. The diversity of respondents by department and position title makes a subtle but systemic issue apparent. Every state organizes and manages its health department differently, presenting a potential obstacle in developing state mandates for sharing data on chronic neurologic disease. In most cases, however, the respondent who completed the assessment was the main point of contact for the surveillance system. Any discussions of chronic neurologic disease surveillance should include individuals from at least both of these disciplines.

The processes for developing chronic neurologic disease surveillance in each state seem to vary considerably. The assessment demonstrated that the association between the number of states that conduct any surveillance for chronic neurologic disease and the number of states in which chronic neurologic diseases are reportable or in which surveillance is legislatively mandated is inconclusive. Though states do not need a legislative mandate or a reporting mandate to conduct surveillance, a number of

states pursue surveillance regardless. This is exemplified by ALS, Parkinson's disease, Alzheimer's disease, Guillain-Barré syndrome, and Creutzfeld-Jakob disease surveillance. Table 2 demonstrates that for each one of these diseases, at least one state conducts surveillance without considering the condition reportable or having a legislative mandate to conduct surveillance. The impetus for states to carry out this surveillance would certainly require further probing.

There were limitations in conducting this assessment. Creutzfeldt-Jakob disease is commonly listed in state's communicable disease reporting laws, not chronic or environmental and therefore, reported to Communicable Disease or Infectious Disease state personnel according to the State Reportable Conditions Assessment (SRCA)<sup>2</sup>. Yet, the responding audience of the assessment was environmental and chronic disease epidemiology points of contact in state health departments. The SRCA was developed by CDC and CSTE to cover state reporting requirements as defined by regulation or legislation, for all conditions defined as reportable by clinicians (health care providers), laboratories, hospitals, and other reporters at the state level. The SRCA demonstrated that 46 states (92%) report Creutzfeldt-Jakob which is significantly more than the 6 that were identified by our environmental and chronic disease points of contact. This gap was exacerbated by the logic model built into the Survey Monkey assessment which directed any responders who selected "no chronic neurologic disease surveillance" to the end of the assessment. Therefore, responders who did not consider Creutzfeldt-Jakob or perhaps Guillain-Barré syndrome as a chronic neurologic disease would not have even received an opportunity to answer. Discrepancies were also found with Parkinson's Disease and Guillain-Barré syndrome. It is likely that information about Creutzfeldt-Jakob, Guillain Barre, and Parkinson's disease surveillance without the consultation of infectious disease epidemiologists was incomplete. (Note: The SRCA did not include ALS in the list of conditions which states were asked to identify as reportable or not.) Further research would be necessary to classify the reporting rules for these conditions to compare with the results from this assessment.

## **References Cited**

1. Survey Monkey (2009) *SurveyMonkey.com*. Portland OR: Ryan Finley.
2. CSTE. State Reportable Conditions Assessment (2009)  
<http://www.cste.org/dnn/ProgramsandActivities/PublicHealthInformatics/StateReportableConditionsQueryResults/tabcid/261/Default.aspx>

## **Appendix A**

ALS State Surveillance Assessment (attached)

## **Appendix B**

### Follow-up Interview Questions

1. Date surveillance started data collection
2. Date surveillance ended data collection
3. What area of the states does the surveillance cover?
  - a. If it isn't the entire state, are there plans to expand the surveillance?
4. What is the legislative mandate for the surveillance?
  - a. Specific law
  - b. General health department authority
  - c. Other
5. What is the case definition?
6. Is reporting active or passive? (please explain)
7. Please describe case reporting including who reports or how cases obtained (e.g., doctors, hospitals, MRI labs, others report, hospital discharge data, mortality data)
8. What variables are you collecting?
9. Do you have a written protocol or plan for the surveillance system?
  - a. Would you be willing to share this with ATSDR?

## Appendix C

Reportability and legislative mandates in states that conduct surveillance for any chronic neurologic disease, by disease and state—ALS State Surveillance Assessment, 2009

State	Alzheimer's disease			Amyotrophic lateral sclerosis			Creutzfeldt-Jakob Disease			Epilepsy			Guillian-Barre Syndrome			Multiple Sclerosis			Parkinson's Disease		
	R	M	S	R	M	S	R	M	S	R	M	S	R	M	S	R	M	S	R	M	S
California			x									x							x	x	x
Delaware							x	x	x				x	x	x						
Idaho							x	x	x						x						
Massachusetts				x	x	x															
Nebraska							x	x	x										x	x	x
New Mexico			x						x										x		x
New York	x	x	x																		
South Carolina			x				x					x			x			x			x
South Dakota			x																		

R, condition is reportable; M, condition is legislatively mandated; S, state conducts surveillance for the condition.