

**Proposal for State and Metropolitan Area-Based ALS
Surveillance**

**Version 6
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INTRODUCTION

Residents of communities living near hazardous waste sites have expressed concerns about elevated rates of selected neurological diseases such as amyotrophic lateral sclerosis (ALS). The absence of population-based surveillance systems or registries for neurological diseases from which estimates of prevalence and incidence could be obtained, as well the complexity in enumerating cases within a specific community, makes it difficult to address these concerns. A bill to amend the Public Health Service Act to provide for the establishment of an Amyotrophic Lateral Sclerosis Registry, S. 1382: ALS Registry Act, was signed into law on October 10, 2008 by President Bush and became Public Law No: 110-373. ATSDR developed and tested methodology for developing a National ALS Registry identifying ALS cases using administrative data from the Centers for Medicaid and Medicare Services (CMS), the Veterans Health Administration (VHA), and the Veterans Benefits Administration (VBA), and a self-registration component because of previous experience trying to obtain information directly from medical care providers, In order to evaluate the completeness of the National ALS Registry, ATSDR is developing 2-3 state-based and 4-6 metropolitan area-based surveillance activities for ALS.

This protocol describes the methodology for developing these state and metropolitan area-based ALS surveillance activities. The primary objective of these surveillance activities is to obtain reliable information on the incidence and prevalence of ALS in a defined geographic area and assess the completeness of the National ALS Registry.

BACKGROUND

Disease Description and Epidemiology

In 1869, the French neurologist Jean-Martin Charcot described a unique condition characterized by deterioration of both lower and upper motor neurons, and this condition was termed amyotrophic lateral sclerosis (ALS).¹ Many people know ALS as Lou Gehrig's disease, named after the famous baseball player who, in 1939, retired because of his illness.

Reports from the United States and other countries indicate an annual incidence rate of 0.2 to 2.4 per 100,000 population and a prevalence of 0.8 to 7.3 per 100,000 population.² The onset of ALS is age-related with the highest rate of onset occurring between 55 and 75 years of age.²⁻⁴ Prognosis also appears to be age-related with slightly better survival occurring among those with a younger age at onset. The average survival time after onset of symptoms is approximately three years, and only a small proportion of patients survive beyond five years.² ALS is more common in males than females by a ratio of 1.5 – 2 to 1,^{4,5} but recent studies have suggested that this sex difference is decreasing over time.^{4,6}

In addition to ALS, several other less common conditions are classified under the general term of motor neuron disease, but ALS accounts for 85 percent or more of all motor neuron cases. Most individuals who are initially diagnosed with these other conditions will ultimately progress to include both upper and lower motor neurons and thus will be diagnosed as having ALS.^{2,7}

Differential diagnosis of ALS requires a neurological exam as well as neurophysiological tests and other tests to rule out non-motor neuron diseases and other motor neuron diseases with restricted presentations. False-negative rates can be high in the early stages of the disease^{8,9} and

false-positive misdiagnoses have been shown to occur in 7 to 8 percent of cases.^{10, 11} The diagnosis of ALS will become more uniform worldwide as the World Federation of Neurology El Escorial criteria and its subsequent revision are utilized.^{12, 13}

Uncertainty about the incidence and prevalence of ALS, as well as the role of the environment in the etiology of ALS, supports the need for a surveillance system for these diseases.^{14, 15} In addition, such an activity could provide an unbiased source from which to recruit patients to participate in future research studies.

Surveillance

Public health surveillance is defined as “the ongoing, systematic collection, analysis, and interpretation of health data essential to the planning, implementation, and evaluation of public health practice, closely integrated with the timely dissemination of these data to those who need to know. The final link of the surveillance chain is the application of these data to prevention and control. A surveillance system includes a functional capacity for data collection, analysis, and dissemination linked to public health programs.”¹⁶ Surveillance is important to monitor changes in incidence and prevalence of a condition. Surveillance data can also be used in planning for health care needs, detecting changes in health practices, and assessing the burden of disease. For chronic diseases, monitoring the burden of disease (morbidity, disability, and mortality) may be very important.¹⁷ To date, national disease surveillance systems have been related primarily to infectious diseases with cancer and birth defects being the two exceptions. In 1992, directors of the World Health Organization (WHO) non-communicable disease collaborating centers and key officials in centers for non-communicable diseases advocated for

the increased surveillance of non-communicable diseases. This recommendation was based on the lack of incidence data for non-communicable diseases.¹⁸

Traditionally, surveillance systems have relied on physicians and other health care providers “reporting” to a specified entity, usually the state or local health department; that information can then be relayed to the next level as appropriate. The designation of “reportable” is conferred by the Council of State and Territorial Epidemiologists (CSTE), which was established in the 1950s. Once a disease has been designated reportable, each state must decide if current health department authorities would include the new disease or whether new legislation must be sought. Historically no non-communicable diseases have been made reportable by CSTE, including cancer. Cancer is reportable in most states; however, this was accomplished by Congress passing Public Law 102-515, the Cancer Registries Amendment Act. This legislation required the authorization of a statewide registry under state law before receiving Federal funds.

Unfortunately, physicians have historically been poor reporters of disease; for that reason laboratory and hospital reporting have been built into surveillance systems.¹⁹ Because physicians do not make good “reporters,” and the history of making diseases nationally reportable has mostly excluded non-communicable diseases, this does not appear to be the best strategy for a national ALS surveillance system. However, it could be an appropriate method for state-based and metropolitan area-based surveillance where it is possible to have direct interaction with medical care providers. State health departments have broad mandates which allow the collection of data for surveillance of diseases affecting their residents. Resources in the form of money and personnel will be provided to the states for this project.

Legislative Mandate

A bill to amend the Public Health Service Act to provide for the establishment of an Amyotrophic Lateral Sclerosis Registry, S. 1382: ALS Registry Act, was signed into law on October 10, 2008 by President Bush and became Public Law No: 110-373. The purpose of the registry as described in the bill, is to: (1) better describe the incidence and prevalence of ALS in the United States; (2) examine appropriate factors, such as environmental and occupational, that might be associated with the disease; (3) better outline key demographic factors (such as age, race or ethnicity, gender, and family history of individuals who are diagnosed with the disease) associated with the disease; and (4) better examine the connection between ALS and other motor neuron disorders that can be confused with ALS, misdiagnosed as ALS, and in some cases progress to ALS. The registry will collect personal health information that may provide a basis for further scientific studies of potential risks for developing ALS.

RATIONALE

State-based and metropolitan area-based surveillance will be conducted to evaluate the completeness of the National ALS Registry. The state-based and metropolitan area-based surveillance activities will rely on obtaining reports from medical providers and abstracting medical records rather than using existing administrative data. The methodology will be similar to that used for state-based cancer registries.

OBJECTIVES

The objective of this project is to develop state-based and metropolitan area-based surveillance activities for ALS. The primary goal of the state-based and metropolitan area-based surveillance

activities is to use these data to evaluate the completeness of the National ALS Registry.

The state and metropolitan areas will be selected to be over representative some racial and ethnicity minorities because of concerns related to possible difference in access to medical care and advocacy organization resources. It is not possible to get an overrepresentation of AI/AN with state populations, however this will be explored as metropolitan areas are added.

PROPOSED SURVEILLANCE DESIGN AND METHODS

A population-based surveillance activity for ALS will be created by identifying persons with ALS from medical care providers. Medical care providers will provide information on or allow abstractors to complete the reporting form for cases of ALS under their care (Attachment 1).

Initially, neurologists and Electromyogram (EMG) laboratories not associated with a neurology practice will be targeted for reporting. Information collected as a part of the abstraction/reporting will include identifying information to be able to de-duplicate records and information on symptoms that will be used to verify the diagnosis. A minimal amount of data will be collected including:

- Name
- Address
- Last 5 digits of the SSN
- Date of birth
- Sex
- Race
- Ethnicity
- Diagnosis
- Date of diagnosis
- Specific diagnosis
- Type of provider making the report
- Payer type

Individuals with an ALS diagnosis as of January 1, 2009 – December 31, 2011 will be included in the surveillance activity. Reporting is expected to begin in March 2011 and continue through

March 2012. Initially reporting will be a mixture of prevalent and incident cases, however as time progresses only incident cases will be reported. State and metropolitan area health departments will publicize the surveillance initiative with providers in their state or metropolitan area through a variety of mechanisms including, but not limited to, neurology meetings, advertisements in neurology publications, and the health department website. The state and metropolitan area health department staff will train medical personnel how to complete the abstract form and assist with abstracting records as requested. To assist with the completing reporting forms, compensation will be available to medical care providers calculated on a per case basis. Physicians will receive \$100 for each case reported to offset reporting costs. An additional \$50 will be available as necessary to offset costs related to medical records abstraction. Each medical provider reporting source should keep a line listing of individuals diagnosed with or thought to have ALS along with information on whether or not the case was reported and if not, the reason. Health department personnel will be in frequent contact with physician's offices and will be available to assist with this activity. Physicians who rely on the health department staff to abstract medical records are not eligible for this compensation.

Demographics of Participant Populations

Currently, three states, Florida, New Jersey, and Texas, and six metropolitan areas, Chicago (Cook and DuPage Counties), Los Angeles, San Francisco (Alameda, Contra Costa, San Francisco, Solano, and San Mateo Counties), Philadelphia (Philadelphia County), Detroit (Wayne County), and Atlanta (Fulton, DeKalb, Cobb, Gwinnett, and Clayton Counties) have been funded to participate in the state-based portion of this surveillance initiative. The racial and ethnic distribution of the states' and metropolitan areas populations shows an over-representation

of African-Americans and Hispanics compared with the United States as a whole.²⁰

Race	US Total Population		All Confirmed Sites	
	%		%	
Total:		308,745,538		80,749,953
White alone	72.4%	223,965,009	64.9%	52,412,676
Black or African American alone	12.6%	38,901,938	15.5%	12,520,709
American Indian and Alaska Native alone	0.9%	2,778,710	0.5%	429,833
Asian alone	4.8%	14,819,786	6.6%	5,359,045
Native Hawaiian and Other Pacific Islander alone	0.2%	617,491	0.1%	105,133
Some other race alone	6.2%	19,142,223	9.3%	7,478,336
Two or more races	2.9%	8,953,621	2.5%	2,055,221
Hispanic or Latino	16.3%	50,325,523	28.7%	23,201,691

We would like to add the metropolitan areas of Baltimore (Howard County, Baltimore County, and Baltimore City) and Las Vegas (Clark County).

Race	US Total Population		Baltimore and Las Vegas	
	%		%	
Total:		308,745,538		3,664,344
White alone	72.4%	223,965,009	56.5%	2,070,650
Black or African American alone	12.6%	38,901,938	23.5%	860,086
American Indian and Alaska Native alone	0.9%	2,778,710	0.6%	20,183
Asian alone	4.8%	14,819,786	7.2%	264,677
Native Hawaiian and Other Pacific Islander alone	0.2%	617,491	0.4%	14,344
Some other race alone	6.2%	19,142,223	8.0%	292,319
Two or more races	2.9%	8,953,621	3.9%	142,085
Hispanic or Latino	16.3%	50,325,523	17.6%	645,068

We anticipate approximately 4500 individuals in the three participating states and 2500 individuals in the metropolitan areas.

Quality Assurance

Quality assurance will be composed of different aspects: the completeness of case ascertainment, the accuracy of the case report, and the accuracy of the diagnosis.

Completeness of case ascertainment

State and local health department staff will be responsible for evaluating the completeness of their case ascertainment prior to submitting their dataset. Death data held by the state or local health departments will be searched for all cause of death codes for ICD-10, G12.2, the code for motor neuron disease (MND) for January 1, 2009 – December 31, 2011. These data will be compared with those obtained from active reporting sources, i.e., reports from medical providers. Because the ICD-10 code is not specific for ALS, the hardcopy death certificate will be examined for anyone not already identified. If the cause of the death is ALS and not one of the other MNDs, the provider signing the death certificate will be contacted to obtain additional information and determine if the individual had ALS between January 1, 2009 and December 31, 2011. If the individual signing the death certificate is a coroner or nursing home, the coroner or nursing home will be contacted to obtain information on the treating physician, if possible, and this individual will be contacted.

Uniform hospital billing data will be searched for ICD-9, 335.20, the specific code for ALS. State health departments will explore the availability of other data sets that can be used for ascertaining missing cases, including but not limited, to advocacy groups (ALS Association, Muscular Dystrophy Association), and pharmacy data for the use of riluzole, the only drug specific for ALS. These data sets will be compared with those cases obtained from active reporting sources. For those cases not already identified, the medical care provider will be contacted to determine why the case was not reported to the health department and to determine if there are additional cases that have not been reported. Staff will be available to train the medical personnel to complete the reporting form and/or abstractors will be available to assist the

medical care provider in completing the reporting forms.

Accuracy of Case Reports

Up to 10% of case reports will be re-abstracted by state health department staff and compared with the original case report. All case reports received will be reviewed for completeness. Physicians' offices will be called to address incomplete forms or other data questions. For physicians/offices with a large number of data questions, state health department staff will re-abstract the case report forms. Discrepancies will be noted and adjudicated. Information about the re-abstractation will be given to the medical providers and state health department staff will provide additional training as needed.

Accuracy of Case Diagnosis

ALS can be difficult to diagnosis because there is no definitive test for the disease. Neurologists do not always agree on the diagnosis, therefore a sample of case reports will be reviewed by the consulting neurologist who is an expert in the diagnosis and treatment of ALS. Completed medical records verification forms will be obtained for at least 10% and not more than 20% of reported cases. Medical providers will be instructed to complete the medical records verification for the selected cases (Attachment 2). This form, along with a copy of the EMG, if available, will be sent to the health department. The health department will check the form for completeness and de-identify the records before forwarding the information to the consulting neurologist. The consulting neurologist will review these records and determine if he agrees with the diagnosis of ALS. To ensure the accuracy of the reporting, the validation will be weighted to review more cases at the beginning of the process and when there are physician

office personnel changes. For cases where the consulting neurologist disagrees with the diagnosis of ALS, the ALS surveillance staff will contact the physician's office and talk with the person completing the form to make sure that the case should have been reported. ALS surveillance staff may go to the provider's office and re-abstract the record. For those cases where the consulting neurologist remains in disagreement with the provider diagnosis, the provider will be contacted to discuss the discrepancy. There will not be a standard data collection for this activity but rather a qualitative discussion between neurologists of the diagnosis. The consulting neurologist will summarize issues related to the discrepancies, such as difficulty applying the El Escorial Criteria. This information will be used to improve training materials and train abstractors.

Human Subjects Protection

ATSDR is requesting a waiver of consent for collecting surveillance data on ALS cases. The research involves no more than minimal risk to the subjects because the information has already been collected as part of the cases' medical care, the waiver will not adversely affect the rights and welfare of the subjects as there is no interaction with the participants; the research could not practicably be carried out without the waiver because of the large number of individuals who would need to be contacted, contact information may not be up-to-date, and many cases may be deceased. In addition, surveillance activities administered by state and metropolitan health departments do not traditionally require consent because of the importance to create an all inclusive unbiased count of those with the disease and requiring consent could result in bias.

The IRB previously granted this project a waiver of consent. In addition to the consent waiver, we are requesting a Waiver of HIPAA Authorization, the project involves minimal risk to the

privacy of the individuals, the information is already available as it was collected as part of medical care and there is no interaction with participants, the waiver will not adversely affect the rights and welfare of the participants, the research could not practicably be carried out without the waiver because of outdated contact information and the large number of people included in the surveillance project. The data security section outlines each entity's plan for protecting the data from improper use and disclosure.

Two health departments have determined that this project is public health surveillance and will collect the information requested without HIPAA Authorization under 45 CFR 164.512 (b)(1)(i) permitted disclosure of PHI by a covered entity to a public health authority without authorization for the purpose of public health surveillance.

This protocol includes surveillance activities within three states, Florida, New Jersey, and Texas. ATSDR would like to add Philadelphia, Detroit, and Atlanta. Data from Philadelphia and Atlanta will be collected by McKing Consulting Staff. The CDC/ATSDR IRB has approved McKing to rely on the CDC/ATSDR IRB. Emory University is a major provider of care to persons with ALS in the Atlanta area. They would like to rely on the CDC/ATSDR IRB. Detroit Department of Health and Wellness Promotion agreed to participate in the project and receives their funding through the fiduciary agency Southeastern Michigan Health Association (SEMHA). Both would like to rely on the CDC/ATSDR IRB.

Historically surveillance activities have been used to identify individuals to ask them to participate in studies; however, there are no plans to do studies at this time. If in the future

studies are planned which involve contacting individuals identified through the surveillance activity to participate in studies, IRB approval will be sought prior to initiation.

Data Security

Health departments will collect the data and transmit the data to ATSDR using a secure file transfer method. Health departments are accustomed to dealing with confidential data collected for surveillance of a variety of diseases including, but not limited to, cancer, HIV, TB, and elevated blood lead levels. An overview of each of the participating state's and ATSDR's data security follows.

Florida

Confidential data are housed on the Florida Department of Health's servers in the department computing center managed by the Division of Information Technology. The department follows industry standard procedures for securing the computers, servers and networks physically and electronically. The computing center is housed in a masonry building with automatic steel doors that are always locked. Physical access to the computing center is restricted to authorized personnel using card-key access. Fire protection is provided by a halon system. Department networks are protected by an internet firewall that requires all of the traffic to pass through a single connection point, thus providing the maximum security possible without restricting legitimate access. Electronic access is restricted by the use of username and password access to authorized personnel only. Passwords need to be of certain minimum length, and has to have combination of alphanumeric and special characters and are changed on a periodic basis. All network and computer systems in the computer room have Uninterruptible Power Supplies (UPS). These ensure continuous service during brief electrical power disruptions and minimize

hardware failures. A diesel generator is available, to provide continuous operations during more extended power failures. All Web interfaces to the project data will employ secure sockets layer security. This means that no clear text will move between client and server.

All department employees/contracted staff must attend security and privacy awareness training prior to accessing information technology resources and/or confidential information. All department employees/contracted staff with access to confidential information must sign and comply with the “Acceptable Use and Confidentiality Agreement” to confirm the individual has read and understands department data confidentiality and security policies.

New Jersey

Confidential public health data are stored, maintained, and backed up on a network of New Jersey Department of Health and Senior Services’ servers. The servers are managed by the NJDHSS Office of Information Technology Services (OITS). The Department network is protected by a three-tiered internet firewall that requires all traffic to pass through a single connection point. Access to the network is restricted by the use of username and password access to authorized personnel only. Passwords need to be of a certain minimum length, composed of alphanumeric and special characters, and must be changed on a periodic basis. PCs and servers are protected with the most up-to-date security patches and antivirus updates.

The server that will house data for the ALS surveillance project is part of the Department’s distributed network. The server is housed in a locked room, and physical access to the room is restricted to authorized personnel using card-key access. The server room has its own dedicated

power supply that ensures continuous service during brief electrical power disruptions. Fire protection is provided by a separate smoke and heat detection system and a halon fire suppression system.

All department employees or contracted staff with access to the network must sign and comply with an “Acceptable Use” policy, to confirm the individual has read and understands department data confidentiality and security policies.

Texas

Confidential data for The Texas Department of State Health Services (DSHS) are managed by the DSHS Information Technology Section. DSHS information security policy establishes Information Resources Architecture Standards relating to Information Security. DSHS contracts with International Business Machines Corporation (IBM) to provide storage of all agency data in their data warehouses. The primary data center is located in San Angelo, Texas and the back-up recovery data center is located in Austin, Texas. The physical security policy and procedures for all Data Centers adhere to those established in the Information Security Controls for State of Texas Data center Services (ISeC) and associated contractual documents. Physical security systems comply with applicable regulations such as building codes and fire regulations. Physical access to information resource facilities is granted only to authorized Users via key-card.

All department employees/contracted staff must complete an approved security awareness training program immediately upon being granted access to any DSHS information resource. All users must sign the Health and Human Services (HHS) Computer Usage Agreement stating they

have read and agree to follow HHS and DSHS requirements regarding computer security policies and procedures.

Detroit

This project is housed in a secure area requiring cardkey for access. Desk top computers are password protected. Data files will be kept on a secure server with other protect health information such as birth and death data, child abuse data, etc. Access to this secure server is limited. Anyone handling data have encrypted drives for data storage and transfer. Each day the ALS surveillance specialist will enter case reports into a secure database. Hardcopy records are kept in a separate office, locked at all times, with limited access. Employees are required to take data security and data sharing policies training. Only staff working on the ALS surveillance project will have access to the data.

Los Angeles and San Francisco Bay Area

Confidential public health data for ALS surveillance are stored, maintained, and backed up on a single server tasked to the California Environmental Health Tracking Program on the Local Area Network (LAN) of the California Department of Public Health (CDPH). This server is managed and maintained by the network administrators of the Division of Environmental and Occupational Disease Control and the Information Technology Services Division of CDPH. This server is housed in a locked room, and physical access to the room is restricted to authorized personnel using card-key access. The server room has its own dedicated power supply that ensures continuous service during brief electrical power disruptions.

The CDPH network is protected by a firewall that requires all traffic to pass through a single connection point via Citrix proxy. Access to the network from outside the LAN is restricted by the use of Citrix key token, username and password by authorized personnel only. Access to the network from within the LAN is restricted by the use of username and password by authorized personnel only. Passwords need to be of a certain minimum length, composed of alphanumeric and special characters, and must be changed on a periodic basis. All PCs and servers on the LAN that might communicate with this server -- as well as the server itself -- are protected with the most up-to-date security patches and antivirus updates.

All department employees or contracted staff with access to the network must pass an annual Information Security Training course.

Atlanta, Philadelphia, Baltimore, and Las Vegas

Data collection in Atlanta, Philadelphia, Baltimore and Las Vegas will be managed by McKing Consulting. McKing will accept case reports through a secure fax which is locked in an office with limited access. Each day the ALS surveillance specialist will enter case reports into a secure database. The database will be maintained on a dedicated computer without Internet Access. The computer will be secured when not in use and the data file will be password protected. The database will be backed-up on a weekly basis to CDC-approved encrypted media which will be kept locked up. Paper files will be kept in a locked file and destroyed at the end of the project. Only staff working on the ALS surveillance project will have access to the data. McKing employees will receive training in the appropriate use and security of PII.

Chicago

Data collection in Cook and Dupage County will be managed by Metropolitan Chicago Healthcare Council-Chicago Hospital Council (MCHC-CHC). MCHC will accept case reports through a secure fax which is locked in an office with limited access. Each day the ALS surveillance specialist will monitor the fax for case reports, put the documents in a folder which will then be entered into a secure database which will be located either in a restricted area or in a private room. Case reports not entered will be kept in a locked file until each individual case is entered. The database will be maintained on a dedicated computer with a privacy screen and without Internet Access. The computer will be secured when not in use and the data file will be password protected. The database will be backed-up on a weekly basis to CDC-approved encrypted media, a thumbprint biometric flash drive, which will also be kept in a locked file. Only the Surveillance Specialist, Project Assistant and Project Manager will have thumbprint access. Paper files will be kept in a locked file and shredded at the end of the project. Only staff working on the ALS surveillance project will have access to the data. Everyone at MCHC working on the ALS Surveillance Project will be certified in HIPAA Privacy Training.

Minimize collection of identifiable information

The information required for reporting a case of ALS to the surveillance activity has been limited to only that needed to describe the basic demographics of the cases being reported and to make sure that an individual truly has ALS and is not already been submitted. To truly de-duplicate case reports, SSN is needed, however only the last five digits SSN will be collected.

ATSDR data management

ATSDR will maintain the National ALS Registry on a secure server or stand-alone hard-drive. Data will be password protected and access to the data will be limited to approved study

personnel. Data from the states and metropolitan areas will be compared with that in the National ALS Registry to evaluate completeness. De-identified data sets will be used for data analysis.

DATA ANALYSIS

Data from the states and metropolitan areas will be compared with the National ALS Registry. Descriptive statistics will be used to describe the differences, if any, between the state/metropolitan area data and the National ALS Registry. This information will be used to identify if there is underreporting in the National ALS Registry for specific groups based on demographics, and to develop strategies to improve the completeness of the National ALS Registry. Although sensitivity and specificity cannot be calculated because the surveillance activities will only report cases of ALS, positive predictive value (PPV) will be calculated. Data from the state and metropolitan areas will be used to calculate incidence and prevalence rates of ALS and to better describe the demographic characteristics (e.g., age, race, sex, and geographic location) of those with ALS. Individual state/metropolitan area data will be analyzed by the respective health departments and not be ATSDR.

CONCLUSION

There is a public health need for accurate estimates of people affected by neurodegenerative diseases to better assess the health care needs of the population, detect changes in health care practices, and assess the burden of disease. This endeavor will provide timely, geographic specific data on ALS which can be used by state health departments to better access the needs of their constituents and will assist ATSDR in evaluating the completeness of the National ALS Registry.

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Attachment 1
ALS Case Reporting Form

Date Completed ___/___/___
Name of person completing the form _____
Job Title _____
Name of Practice _____
Phone number (____) _____ - _____

ALS Case Reporting Form

This form should only be completed for individuals meeting the El Escorial Criteria for diagnosing ALS including definite, probable, and possible ALS. The diagnosis of ALS requires the presence of each of the following:

1. Lower Motor Neuron signs (by clinical, electrophysiological, or neuropathological examination) in 1 or more of 4 regions (bulbar, cervical, thoracic, and lumbosacral). Signs of lower motor neuron degeneration include: weakness, muscle atrophy and fasciculations.
2. Upper Motor Neuron signs (by clinical examination) in 1 or more of the 4 regions. Signs of upper motor neuron degeneration include: slowed movements, increased muscle tone or spasticity, spastic gait.
3. Progression of signs within a region or to other regions

Definite ALS = Upper Motor Neuron + Lower Motor Neuron signs in 3 regions

Probable ALS = Upper Motor Neuron + Lower Motor Neuron signs in 2 regions with Upper Motor Neuron signs rostral to Lower Motor Neuron signs

Probable ALS, lab supported = Upper Motor Neuron + Lower Motor neuron signs in 1 region with evidence by EMG of lower motor neuron involvement in another region.

Possible ALS = Upper Motor Neuron + Lower Motor Neuron signs in 1 region or Upper Motor Neuron signs in 2 or 3 regions, such as monomelic ALS, progressive bulbar palsy, and primary lateral sclerosis

Demographic Information

1. Subject Name:
 1. Last Name _____
 2. First Name _____
 3. Middle Name or Initial _____
 4. Suffix _____
2. Address:
 1. Number _____
 2. Street _____
 3. City _____
 4. State _____
 5. Zip Code _____
3. Social Security Number (last 5 digits only) __ - ____ - ____
4. Date of Birth: ___/___/____ (mm/dd/yyyy)
5. Sex: Male Female
6. Race (as reported by subject – check all that apply):
 - Asian
 - Black/African American
 - White
 - Unknown
 - Other: _____
7. Ethnicity:
 - Hispanic or Latino
 - Non Hispanic or Latino
 - Unknown

8. Country of Birth: _____

Diagnosis Information

9. El Escorial Criteria as determined by an ALS specialist (check one)

- Definite
- Probable
- Probable (lab supported)
- Possible
- Not Classifiable

10. Month/Year of Diagnosis ___/___ (mm/yyyy)

11. Month/Year of Onset of Symptoms ___/___ (mm/yyyy)

12. Provider Making the Report

- Neurologist (ALS specialist)
- Neurologist (other)
- Psychiatrist
- Family/Internal Medicine/General Practice

13. Does the patient have dementia diagnosed by a neurologist?

- Yes
- No
- Don't know

14. Does the patient have an immediate family member (parent, sibling, child) who has/had ALS?

- Yes
- No
- Don't know

15. Payer Type

- Medicare
- Medicaid
- HMO
- Private Insurance
- Self-pay
- Veterans Administration
- Other

Attachment 2

ATSDR AMYOTROPHIC LATERAL SCLEROSIS
MEDICAL RECORD VERIFICATION FORM

Instructions for Completing the Medical Record Verification Form

Please complete the form attached for each participant selected by looking at the 1st and the last neurology note. If you are unable to complete the form with just two notes, please review the rest of the record. Each question should be answered. For questions that have multiple subquestions, such as muscle atrophy, please continue reviewing the medical record until you can answer at least one of the subquestions (tongue, upper extremity, lower extremity, or unspecified location).

Abstractor (Name)
Site Specific Subject ID:

Abstraction Date ___/___/___

**ATSDR AMYOTROPHIC LATERAL SCLEROSIS MEDICAL RECORD
VERIFICATION FORM**

1. Difficulty swallowing (dysphagia) (at any time): Yes No or not noted
2. Difficulty talking (dysarthria) (at any time): Yes No or not noted
3. Limb weakness (at any time):
 - A. Upper extremity Yes No or not noted
 - B. Lower extremity Yes No or not noted
 - C. Generalized Yes No or not noted
4. Hyper-active Reflexes (at any time)
 - A. Upper extremity (Biceps, Brachioradialis or Triceps)
 Yes No or not noted
 - B. Lower extremity (Knee jerk, ankle jerk or positive Babinski response)
 Yes No or not noted
5. Fasciculations (at any time)
 - A. Tongue Yes No or not noted
 - B. Upper extremity Yes No or not noted
 - C. Lower extremity Yes No or not noted
 - D. Chest Yes No or not noted
 - E. Unspecified location Yes No or not noted
6. Muscle atrophy (at any time)
 - A. Tongue Yes No or not noted
 - B. Upper extremity Yes No or not noted
 - C. Lower extremity Yes No or not noted
 - D. Unspecified location Yes No or not noted
7. Site of Onset of Weakness (initial visit only, check one):
 Bulbar Truncal Generalized Respiratory
 Limb Upper Limb Lower None Unknown
8. Ever treated with riluzole (at any time):
 Yes No
9. Date of Death (if applicable and known): / (mm/yyyy) NA Don't know
10. **Please attach a copy of the most recent EMG report to this abstraction form.**
 Yes, attached No, not available

For Official Use Only

1 2 3 4 5

