Proposal for State and Metropolitan Area-Based ALS Surveillance

Version 2
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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 secondary goal of the surveillance activities is to obtain reliable and timely information on the incidence and prevalence of ALS and to better describe the demographic characteristics (e.g., age, race, sex, and geographic location) of those with ALS. 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. 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
Date of diagnosis
Specific diagnosis
Type of provider making the report

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 $25-$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.

Demographics of Participant Populations

Currently, three states, Florida, New Jersey, and Texas, have been funded to participate in the state-based portion of this surveillance initiative. The racial distribution of the states' populations is similar to the United States as a whole.[20]

Race	US Total Population		Texas, Florida, New Jersey	
Total:	**%**	**301,237,703**	**%**	**50,686,978**
White alone	74.3%	223,965,009	73.1%	37,040,347
Black or African American alone	12.3%	37,131,771	13.2%	6,688,461

American Indian and Alaska Native alone	0.8%	2,419,895	0.4%	190,589
Asian alone	4.4%	13,164,169	3.7%	1,860,507
Native Hawaiian and Other Pacific Islander alone	0.1%	446,164	0.1%	31,043
Some other race alone	5.8%	17,538,990	7.8%	3,955,077
Two or more races	2.2%	6,571,705	1.8%	920,954

The ethnic distribution shows these three states have a significantly higher number of individuals identified as Hispanic or Latino.20

Ethnicity	**United States**		**Florida, New Jersey, Texas**	
	Estimate		Estimate	
Total:	301,237,703	%	50,686,978	%
Not Hispanic or Latino	255,805,545	84.92%	37,014,489	73.03%
Hispanic or Latino:	45,432,158	15.08%	13,672,489	26.97%
Mexican	29,318,971	9.73%	8,072,706	15.93%
Puerto Rican	4,127,728	1.37%	1,217,529	2.40%
Cuban	1,572,138	0.52%	1,197,907	2.36%
Dominican (Dominican Republic)	1,249,471	0.41%	322,529	0.64%
Central American	3,592,810	1.19%	951,673	1.88%
South American	2,544,070	0.84%	1,031,062	2.03%
Other Hispanic or Latino	3,026,970	1.00%	879,083	1.73%

Additional metropolitan areas might be added, with IRB approval, to increase the numbers of other racial minorities covered by the surveillance activities. We anticipate approximately 4500 individuals in the three participating states.

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

On a yearly basis, death data 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.

Quarterly 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

A 10% sample of case reports will be re-abstracted by state health department staff and compared with the original case report. Discrepancies will be noted and adjudicated. The

information on the re-abstraction 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. 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.

This protocol includes surveillance activities within three states, Florida, New Jersey, and Texas. If ATSDR decides to expand the protocol to additional states and/or metropolitan areas to increase the representation of racial and ethnic minorities, IRB approval will be sought via an amendment to this protocol.

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.

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 included: 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):
    - ☐ American Indian/Alaska Native
    - ☐ Asian
    - ☐ Native Hawaiian/Other Pacific Islander
    - ☐ Black/African American
    - ☐ White
    - ☐ Unknown
    - ☐ Other:_____
7. Ethnicity:
    - ☐ Hispanic or Latino
    - ☐ Non Hispanic or Latino

OVER ➡

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)
    - ☐ Physiatrist
    - ☐ 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

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) Abstraction Date __ __/__ __/__ __
Site Specific Subject ID:

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

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