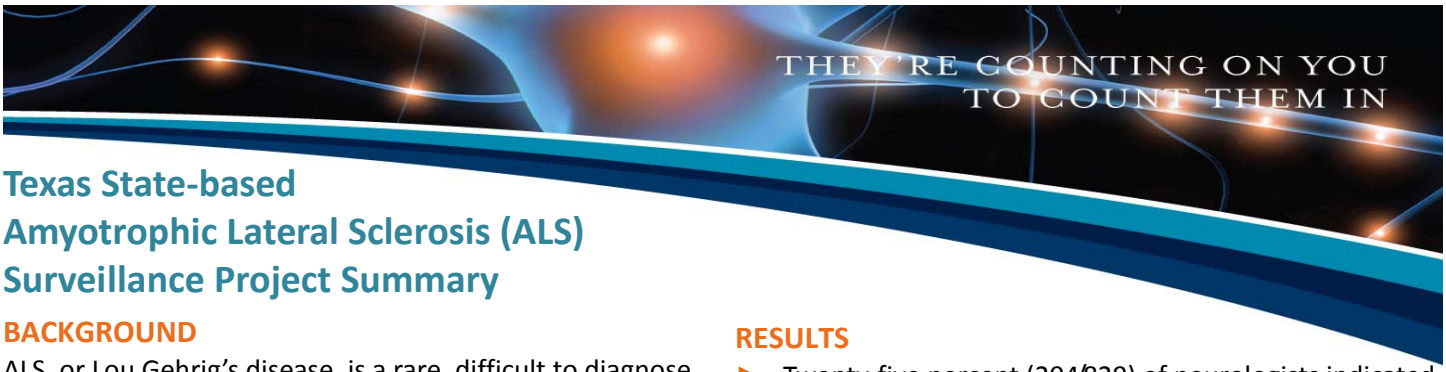

Texas State-based Amyotrophic Lateral Sclerosis (ALS) Surveillance Project Summary

BACKGROUND

ALS, or Lou Gehrig's disease, is a rare, difficult to diagnose neurological condition with no known cause or cure. Because ALS is a non-notifiable disease, little is known about its incidence and prevalence in the U.S. To help learn more about ALS, the federal Agency for Toxic Substances and Disease Registry (ATSDR) maintains the National ALS Registry (Registry).[1,2] The Registry identifies ALS cases using national administrative databases, including those from Medicaid, Medicare, and the Veterans Health Administration and Veterans Benefits Administration, and by patient self-enrollment through a web portal. ATSDR funded McKing Consulting Corporation (McKing) to complete surveillance projects to gather reliable and timely data to describe the incidence and demographic characteristics of ALS and to assist ATSDR in evaluating the completeness of the Registry. Surveillance projects were conducted in three states (Florida, New Jersey, and Texas) and in eight metropolitan areas (Atlanta, Baltimore, Chicago, Detroit, Las Vegas, Los Angeles, Philadelphia, and San Francisco). This summary describes the Texas project.

METHODS

McKing partnered with the Texas Department of State Health Services (DSHS) to conduct the project. All neurologists practicing in the state of Texas were asked if they diagnosed or provided care for ALS patients. Emphasis was placed on neurologists specializing in the diagnosis/care of persons with ALS who practice at referral centers that typically see more than 50 patients per year. Neurologists were asked to submit one-page case reports for ALS patients under the doctor's care who were alive at some point between January 1, 2009 and December 31, 2011. A medical record verification form (MRVF) and an electromyogram (EMG) report were requested for a sample of cases and reviewed by an independent consulting neurologist to confirm ALS diagnosis. Death data were reviewed to identify additional cases, and attempts were made to obtain case reports for decedents that were not already reported. Compensation was offered to neurologists for completed forms. No patients were contacted. This project was approved by the Centers for Disease Control and Prevention Institutional Review Board (IRB) and the Texas DSHS IRB.

RESULTS

- Twenty-five percent (204/829) of neurologists indicated that they diagnosed and/or cared for ALS patients and 54% (110/204) of those neurologists reported cases (Table 1).

Table 1: Recruitment and Participation of Neurologists in Texas

	n	%*
All neurologists	829	100.0
Diagnosed/care for ALS patients in reporting period	204	24.6
Reported cases	110	13.3
Did not report cases	94	11.3
Diagnose/care for ALS patients, not in reporting period	67	8.1
Will not diagnose/care for ALS patients	503	60.7
Unknown	54	6.5
Other physicians reporting cases	1	n/a

*Does not add up to 100% due to rounding.

- A total of 1,670 case reports were received; 1,423 were unique cases.
- Ninety-two percent (1,305/1,423) of cases were reported as "definite," "probable," or "probable-lab supported" according to the El Escorial criteria.[3] Ninety-four percent (217/232) of the requested MRVFs were received; 76% (165/217) were classified as "definite," "probable," or "probable-lab supported," and 24% (52/216) were classified as "possible" or "not classifiable" by the consulting neurologist.
- Seventy-nine percent of cases were 50 years of age or older at diagnosis, 56% were male, 80% were white, and 74% were not Hispanic or Latino (Table 2).
- Of the 1,245 cases for whom data were available, 50% had symptoms for 12 months or less before diagnosis. Ninety percent of the 1,245 cases were diagnosed within 36 months of having symptoms.
- Forty percent (563/1,423) of cases had only federal payers [Medicare, Medicaid, Veterans Affairs (VA)], 39% (554/1,423) had only non-federal payers (HMO, private insurance, self-pay, or other), and 21% (306/1,423) had both federal and non-federal payers.

Table 2: Demographic Characteristics of All Reported ALS Cases in Texas, n=1,423		
Demographic Characteristic	**n**	**%***
Age (years)		
Under 40	82	5.7
40 – 49	201	14.1
50 – 59	351	24.7
60 – 69	426	29.9
70 – 79	276	19.4
80 or older	76	5.3
Unknown	11	0.8
Sex		
Male	796	55.9
Female	627	44.1
Race		
White alone	1135	79.8
Black/African American alone	93	6.5
Asian alone	21	1.5
Other**	1	0.0
Unknown	173	12.2
Ethnicity		
Hispanic	213	15.0
Not Hispanic or Latino	1053	74.0
Unknown	157	11.0

*May not add up to 100% due to rounding.
**Those with multiple races are listed here.

DISCUSSION

- One-quarter of neurologists diagnosed or cared for patients with ALS during the reporting period and a little more than one-half of them reported cases.
- Most of the ALS referral centers in the state participated and submitted the majority of case reports.
- Some non-referral center practices and the VA hospitals and clinics had cases to report but declined to participate. However, it is unclear if providers at these practices would have reported unique ALS cases.
- Many unique names were identified in the death data that were not reported to the project. It is unknown if any of these individuals were true ALS cases.
- The expected number of cases in the three-year period was 2,012[4,5] and only 1,423 cases were reported to the project. Incidence rates were not calculated because they may not yield accurate estimates of the rates due to underreporting.
- The geographic distribution of ALS cases across the state reflected the Texas population distribution with approximately two-thirds of reported cases residing in Texas' main metropolitan areas (Austin, Houston, Dallas, and San Antonio).
- Although the major metropolitan areas have large specialty centers, there are many rural providers spread throughout the state who each see a small number of patients. It may be difficult for persons with ALS to travel from remote rural locations to the metropolitan areas.
- Lessons learned regarding physician recruitment that may prove helpful in replicating similar surveillance efforts include the following:
  - Creating targeted phone call lists, sending faxes immediately followed by phone calls, and conducting on-site visits appeared to increase visibility of the project, but did not seem to increase participation of non-referral center sites.
  - Garnering the support and participation of major ALS referral centers in the catchment area was imperative.
- The results of this project demonstrate that active case ascertainment is expensive and laborious. In addition, not all neurologists are willing to provide case reports, which supports the methodology used by the National ALS Registry.

FOR MORE INFORMATION
PLEASE VISIT THE ATSDR WEB SITE:
HTTP://WWWN.CDC.GOV/ALS/ALSSTATEMETRO.ASPX

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