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Updated Prevalence and Demographic Characteristics for ALS Cases in Texas, 2009–2011

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Abstract

Objectives—Amyotrophic lateral sclerosis (ALS) is a rare motor neuron disease with incidence rates ranging from 1 to 2/100,000 person-years. The Texas Department of State Health Services previously conducted surveillance for ALS in three metropolitan areas of Texas. This project provides an update to this research, while expanding its scope to the entire state.

Methods—The Texas Department of State Health Services contacted neurologists throughout Texas to determine whether they diagnose or treat patients with ALS. Those neurologists who cared for Texas residents with ALS between 2009 and 2011 were asked to complete a one-page case reporting form for each patient.

Results—A total of 1422 unique cases were received. The average crude annual incidence rate was 1.30/100,000 person-years and the 2009 period prevalence rate was 2.92/100,000 individuals. Reported cases were most likely to be 60 to 69 years old, non-Hispanic, white, and men.

Conclusions—This project provides an update to previously published Texas-specific epidemiological data regarding ALS; also, we note that our findings are consistent with previously published studies.

Keywords

amyotrophic lateral sclerosis; epidemiology; prevalence; surveillance

Amyotrophic lateral sclerosis (ALS) is a motor neuron disease that affects both upper and lower motor neurons. The literature describes incidence rates ranging from 1 to 2/100,000 person-years.[1] It is the most common motor neuron disease and most often presents later in life.[2–4] The incidence of ALS has been reported to dramatically increase in people aged 60 years to their early 70s and begins to decline in people in their late 70s.[1,5] In addition, a predominance of male, white, and non-Hispanic cases have been reported.[6–8] The Texas

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Department of State Health Services (DSHS) previously conducted surveillance for ALS in three metropolitan areas of Texas (San Antonio, El Paso, and Lubbock) from 1998 to 2003.[9] Wagner et al[9] reported point prevalence estimates for each of the three county areas (range 1.0–2.2/100,000) as well as for the overall geographic area (1.6/100,000). This article provides an update on ALS in the entire state of Texas for the period 2009–2011.

The National ALS Registry identifies individuals with ALS from patient self-enrollment and through administrative databases, including Medicare, Medicaid, the Veterans Benefits Administration, and the Veterans Health Administration.[10] To validate the National ALS Registry, several active case surveillance projects were initiated in state and metropolitan areas. To conduct this surveillance DSHS collected case reports from Texas neurologists who indicated that they had diagnosed or cared for patients with ALS from January 1, 2009 through December 31, 2011. This report describes those findings and explains how they contribute to the understanding of ALS in Texas.

Methods

Neurologist Identification and Recruitment

DSHS obtained a list of neurologists practicing in 12 sub-specialties from the Texas Medical Board who received a letter outlining the project. Neurologists in specialties unlikely to diagnose or care for patients with ALS (eg, neuroradiologists, neuropathologists, pediatric neurologists) were later removed from the list for further contact. DSHS staff followed up the remaining neurologists with additional letters and telephone calls to determine whether they diagnosed or provided care to patients with ALS during the surveillance period (2009–2011).

Data Collection

Neurologists who diagnosed or provided care to patients with ALS were asked to submit case reports for eligible cases, which included those who were under a doctor's care at any time between January 1, 2009 and December 31, 2011, met one of the El Escorial criteria,[11] and had a Texas address during the reporting period. Because there is no definitive test for ALS, the El Escorial criteria were created to assist in categorizing patients in clinical trials and to aid in the diagnosis of ALS. The diagnosis of ALS relies on the results of a clinical examination and history in combination with electromyograms (EMGs) to detect signs of upper and/or lower motor neuron degeneration. Patients are classified on a spectrum ranging from "possible ALS" to "definite ALS" based on symptoms and other criteria (Table 1). For this project, those who met criteria to be classified as clinically possible or higher were included as cases of ALS. Neurologists used the project's case reporting form to provide information regarding patient demographic characteristics and diagnostic data such as the El Escorial[11] classification level and dates of symptom onset and diagnosis. All of the forms received by DSHS were compiled into a patient database in which all duplicate reports were then removed by McKing Consulting Corporation, the firm that performed this task for all project sites, to maintain consistency. To ensure the accuracy of diagnosis, a sample of cases was selected for verification. The neurologist was asked to complete a medical record verification form (MRVF) for each selected case, and to attach an EMG report when

available. The project followed the Centers for Disease Control and Prevention institutional review board–approved ALS Surveillance project protocol. The project also was approved by the Texas DSHS institutional review board. No patients were contacted.

Data Analysis

Crude annual incidence rates were calculated using the number of cases diagnosed in 2009–2011 and the 2010 US Census population data as the denominator.[12,13] Age-adjusted average annual incidence rates by sex were standardized to the year 2000 US Standard Population.[11] All of the incidence rates are presented as cases per 100,000 person-years. Period prevalence for 2009 was calculated by using all of the cases diagnosed before 2010 and the 2010 US Census Population estimate for Texas (population 25,145,561). Data were analyzed using Microsoft Excel (Microsoft, Redmond, WA) and SPSS 19.0 (IBM SPSS Statistics, Armonk NY).

Results

Neurologists

A total of 829 neurologists, those in specialties most likely to have cared for patients with ALS, were contacted to determine whether they diagnose or care for patients with ALS. Overall, 61% of the neurologists (503/829) said they would not diagnose or treat patients with ALS. Eight percent of the neurologists (67/829) reported they would diagnose or care for patients with ALS but did not have eligible cases to report, and 6% (54/829) did not provide information to the project. A total of 25% of the neurologists (204/829) diagnosed or provided care to patients with ALS in the project's time period (2009–2011), of whom 54% (110/204) reported eligible cases.

Case Reporting

Overall, 1670 case reports were collected, and after removing duplicate cases, a total of 1422 unique cases remained; this is 71% of the 2012 expected cases based on the 2010 Texas population and national incidence and prevalence estimates.[1,12,13] The geographic distribution of ALS cases across the state closely mirrored that of the known population density, with the majority of reported patients residing in Texas' three main metropolitan areas: Houston, Dallas, and San Antonio. Five major ALS treatment centers for Texas, located in the aforementioned cities, reported 71.5% of total number of cases received during the project.

Incidence and Prevalence

A total of 976 incident cases were diagnosed in 2009, 2010, and 2011 (Table 2). Combined, the crude annual incidence rates for 2009, 2010, and 2011 were 1.17, 1.30, and 1.41/100,000 person-years, respectively, and the average crude annual incidence rate was 1.29/100,000 person-years. In general, age-specific average annual incidence rates increased with age until 69 years or older. A total of 733 reported cases were diagnosed before 2010, thus the 2009 period prevalence rate was approximately 2.92/100,000.

Demographic Distribution of Cases

Reported cases were most likely to be older, non-Hispanic, white, and men. More than half of the reported cases were 50 to 69 years of age, with approximately one-fourth above and one-fourth below that age range. According to 2010 US Census data for Texas, 50.4% of the population is female[12,13]; however, of the ALS cases reported, males had a slightly higher prevalence (55.9%). Among all of the reported cases (N = 1422) 80% were white, 7% were black/African American, and 1% were Asian. With respect to ethnicity, 15% were Hispanic (Table 3).[14]

Case Validation

An MRVF and EMG report were requested for a sample of case reports. A total of 232 MRVFs representing 14% of the 1670 collected case reports were requested, and 217 (94%) of them were received. The consulting neurologist determined all but five (2.3%) of these cases to be ALS based on the El Escorial criteria[11] and they were removed from the dataset.

Discussion

This study provides updated information to previous descriptions of the ALS disease burden in Texas. As with previous Texas-based studies and other reported literature,[1,9,15] we found cases were more likely to be white, older, and non-Hispanic. Previous prevalence estimates of ALS in Texas[9] were lower than those from this surveillance project (1.3/100,000 vs 2.92/100,000, respectively); however, those estimates were of point prevalence, the prevalence rate at a single point in time, rather than period prevalence, the prevalence rate for the entire year. In addition, the previously reported rates represent only three geographic areas in Texas, whereas these estimates represent the entire state.

Only 54% of neurologists who diagnosed or cared for patients with ALS in the reporting period actually reported cases to the project which resulted in underreporting of cases. Based on the Texas population and national incidence and prevalence, we estimate that 71% of the expected cases were reported. In addition, most of the cases were reported from the ALS treatment centers located in metropolitan areas. As such, we believe the incidence and prevalence rates found in this project are lower than the true rates and may underrepresent cases in more rural areas. Despite the apparent underreporting, we believe the trends in demographics are correct because they concur with the existing literature. The number of diagnosed cases begins to increase significantly at age 40 and the majority of cases (54.8%) were diagnosed in patients between the ages of 50 and 70. We found a slightly higher prevalence of ALS reported in men versus women. As the literature suggests, the reported cases reflect a much higher prevalence in white patients than other races, with a ratio of almost 4:1.[15] The majority of patients also were reported to be of non-Hispanic ethnicity.[16]

Although there are limitations to this study, it is an important update to previously published findings. Updating previously published incidence and demographic characteristics will help to clarify the numbers of individuals with ALS in Texas and assist medical providers and healthcare decision-makers in better planning for their health care needs. Although there was

a lack of participation by many neurologists treating patients with ALS, this was the first statewide effort in Texas to conduct active ALS surveillance.

Conclusions

Our findings are consistent with current trends in the literature and provide information for describing the characteristics of people with ALS in Texas. In addition, through our case verification process, we are confident that the cases of ALS reported to our project were actual ALS cases. Future projects examining the incidence of ALS in Texas may benefit from using additional case finding techniques such as the use of hospital and death certificate data in conjunction with active surveillance to capture all cases of ALS.

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Key Points

- The average crude annual incidence rate of amyotrophic lateral sclerosis was 1.30/100,000 person-years and the 2009 period prevalence rate was 2.92/100,000 individuals.
- Reported cases were most likely to be 60 to 69 years old, non-Hispanic, white, and men.
- The reported incidence and prevalence rates of amyotrophic lateral sclerosis are consistent with previously published studies.

Table 1

El Escorial criteria

Definite ALS	Presence of upper motor neuron and lower motor neuron signs in the bulbar region and at two of the other spinal regions
Probable ALS	Presence of upper motor neuron and lower motor neuron signs in at least two regions with upper motor neuron signs rostral to lower motor neuron signs
Probable ALS, laboratory results supported	Presence of upper motor neuron and lower motor neuron signs in one region with evidence by EMG of lower motor neuron involvement in another region
Possible ALS	Presence of upper motor neuron and lower motor neuron signs in one region or upper motor neuron signs in two or three regions, such as monomelic ALS, progressive bulbar palsy, and primary lateral sclerosis

ALS, amyotrophic lateral sclerosis; EMG, electromyography.

Table 2

Annual incidence rates for ALS diagnosed in Texas, 2009–2011 (N = 976)

Time period	Cases, n	Incidence rates (per 100,000 person-years)	95% CI
2009	294	1.17	1.04–1.30
2010	327	1.30	1.16–1.44
2011	355	1.41	1.26–1.56
Overall	976	1.29	1.15–1.43

ALS, amyotrophic lateral sclerosis; CI, confidence interval.

Table 3

Demographic characteristics for reported ALS cases diagnosed or receiving care in Texas, 2009–2011

	Texas ALS cases (N = 1422)		Overall Texas population[16] (N = 25,145,561)	
	n	%[a]	n	%[a]
Age, y				
<30	16	1.1	11,291,832	44.9
30–39	66	4.6	3,524,021	14.0
40–49	201	14.1	3,455,262	13.7
50–59	350	24.6	3,097,793	12.3
60–69	426	30.0	2,027,867	8.1
70–79	276	19.4	1,096,401	4.4
≥80	76	5.3	652,385	2.6
Unknown	11	0.8	0	0.0
Sex				
Male	795	55.9	12,472,280	49.6
Female	627	44.1	12,673,281	50.4
Race				
Asian	21	1.5	964,596	3.8
Black/African American	93	6.5	2,979,598	11.8
White	1134	79.7	17,701,552	70.4
Unknown	173	12.2	0	0.0
Other	1	0.0	3,499,815	14.0
Ethnicity				
Hispanic or Latino	213	15.0	9,460,921	37.6
Non-Hispanic or Latino	1053	74.0	15,684,640	62.4
Unknown	156	11.0	0	0.0
Total	1422	100.0	25,145,561	100.0

[a] May not add to 100.0% because of rounding.

ALS, amyotrophic lateral sclerosis.