Las Vegas Metropolitan Area 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 Las Vegas project.

METHODS

All neurologists practicing in Clark County, Nevada, as well as neurologists specializing in the diagnosis/care of persons with ALS practicing at five referral centers in Los Angeles, California, four referral centers in Arizona, and one referral center in Utah that typically see more than 50 patients per year were asked if they diagnosed and/or provided care for ALS patients that resided in Clark County. 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.

RESULTS

- Twenty-six percent (20/77) of neurologists indicated that they diagnosed and/or cared for ALS patients and 40% (8/20) of those neurologists reported cases (Table 1). All major referral centers in the region participated.

Table 1: Recruitment and Participation of Neurologists

	n	%*
All neurologists	77	100.0
Diagnosed/care for ALS patients in reporting period	20	26.0
Reported cases	8	10.4
Did not report cases	12	15.6
Diagnose/care for ALS patients, not in reporting period	26	33.8
Will not diagnose/care for ALS patients	31	40.3

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

- A total of 95 case reports were received; 85 were unique cases.
- Ninety-eight percent (83/85) of cases were reported as "definite," "probable," or "probable-lab supported" according to the El Escorial criteria.[3] All 17 requested MRVFs were received; 88% (15/17) were classified as "definite," "probable," or "probable-lab supported" and 12% (2/17) were classified as "possible" by the consulting neurologist.
- Eighty-four percent of cases were 50 years of age or older at diagnosis, 52% were male, 83% were white, and 79% were not Hispanic or Latino (Table 2).
- Of the 73 cases for whom data were available, 50% had symptoms for 12 months or less before diagnosis. Ninety percent of the 73 cases were diagnosed within 37 months of having symptoms.

- Forty-seven percent (40/85) of cases had only federal payers [(Medicare, Medicaid, Veterans Affairs (VA)], 31% (26/85) had only non-federal payers (HMO, private insurance, self-pay, or other), and 22% (19/85) had both federal and non-federal payers.

Table 2: Demographic Characteristics of All Reported ALS Cases in Clark County, Nevada, n=85

Demographic Characteristic	n	%*
Age (years)		
Under 40	2	2.4
40 – 49	12	14.1
50 – 59	18	21.2
60 – 69	22	25.9
70 – 79	24	28.2
80 or older	3	3.5
Unknown	4	4.7
Sex		
Male	44	51.8
Female	41	48.2
Race**		
White alone	73	85.9
Black/African American alone	6	7.1
Asian alone	3	3.5
Unknown	3	3.5
Ethnicity		
Hispanic	8	9.4
Not Hispanic or Latino	67	78.8
Unknown	10	11.8

*May not add up to 100% due to rounding.
**There were no cases reported with multiple races.

DISCUSSION

- One quarter of neurologists diagnosed or cared for patients with ALS during the reporting period and 40% of them reported cases.
- All ALS referral centers in the region participated. Some non-referral center practices in the region participated. Some non-referral center practices had cases to report, but declined to participate. However, it is unclear if providers at these practices would have reported unique ALS cases.
- Thirty-two 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 156[4,5] and only 85 cases were reported to the project. Incidence rates were not calculated because they may not yield accurate estimates of the rates due to underreporting.
- 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 practices.
  - Garnering the support and participation of major ALS referral centers in the catchment area was imperative.
  - Unique cases were reported by referral centers in Los Angeles, California and Scottsdale, Arizona suggesting that patients residing in Clark County, Nevada travel long distances for diagnosis and/or care.
- 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

REFERENCES

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