Los Angeles and San Francisco Bay 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.[1,2] 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 Los Angeles (LA) and San Francisco Bay Area (SFBA) projects.

METHODS

McKing partnered with the California Environmental Health Tracking Program (CEHTP), located in the California Department of Public Health, to conduct this project. All neurologists practicing in Los Angeles County and five San Francisco Bay Area counties: San Francisco, Alameda, Contra Costa, San Mateo and Solano, California were asked if they diagnosed or provided care for ALS patients who resided in these areas. 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 (CDC) Institutional Review Board.

RESULTS

- Nine percent (93/1,056) of neurologists indicated that they diagnosed and/or cared for ALS patients and 83% (77/93) of those neurologists reported cases (Table 1).
- All 12 of the ALS care centers and large practices in both regions participated in the project.

Table 1: Recruitment and Participation of Neurologists in LA and SFBA

	LA		SFBA	
	n	%	n	%
All neurologists	675	100.0	381	100.0
Diagnosed/cared for ALS patients in reporting period	42	6.2	51	13.4
Reported cases	30	4.4	47	12.3
Did not report cases	12	1.8	4	1.0
Diagnose/care for ALS patients, not in reporting period	72	10.7	23	6.0
Will not diagnose/care for ALS patients	553	81.9	291	76.4
Unknown	8	1.2	16	4.2
Other physicians reporting cases*	0	--	1	--

*Not included in the total.

- Using 2010 U.S. Census population data and estimates of incidence and prevalence, we expected to identify 1,145 unique cases; 360 and SFBA and 785 in LA.[3,4] A total of 1,085 unique cases, which is 150% (540/360) of the expected cases in SFBA and 69% (545/785) of the expected cases in LA.
- The overall age-adjusted incidence rate was 1.7 per 100,000. The age-adjusted incidence estimate was higher in SFBA at 2.0 per 100,000 compared with LA at 1.2 per 100,000.
- Males represented 57% of cases; 59% and 55% in SFBA and LA, respectively. White cases made up 72% in SFBA and 70% in LA. Both sites had substantial yet differing proportions of cases with race or ethnicity not reported (Table 2).
- There were more foreign-born cases in LA (22%) versus SFBA (14%). County of birth was missing in 25% of cases.
- Mean age at diagnosis was 61.5 years (62.2 years in SFBA and 60.8 in LA). The mean age of symptom onset was 59.7 years with a significant difference across the two sites, 60.5 in SFBA and 58.8 in LA.
- For both sites, 78% of cases were reported as "definite," "probable," or "probable-lab supported" using the El Escorial criteria.[5]

Table 2: Demographic Characteristics of All Reported ALS Cases in LA and SFBA, n=1,085

	LA		SFBA	
Demographic Characteristic	**n**	**%***	**n**	**%***
Age (years)				
Under 30	10	1.8	5	0.9
30 – 39	29	5.3	19	3.5
40 – 49	69	12.7	66	12.2
50 – 59	124	22.8	134	24.8
60 – 69	151	27.7	153	28.4
70 – 79	105	19.3	113	20.9
80 or older	45	8.2	47	8.7
Unknown	12	2.2	3	0.6
Sex				
Male	302	55.4	321	59.4
Female	243	44.6	219	40.6
Race				
Asian	46	8.4	64	11.9
Black/African American	55	10.1	34	6.3
White	379	69.5	390	72.2
Unknown	65	11.9	52	9.6
Other	--	--	9	1.7
Ethnicity				
Hispanic	102	18.7	56	10.4
Not Hispanic or Latino	314	57.6	446	82.6
Unknown	129	23.7	38	7.0

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

DISCUSSION

- ALS surveillance in two ethnically diverse California metropolitan areas successfully identified at least 94% of expected cases.
  - Many fewer cases than expected were reported in LA despite extensive outreach efforts, while efforts in SFBA yielded a greater than expected number.
  - Variations in the number of reported cases may be explained by racial and ethnic differences in the two regions or other factors like access to care, referral patterns, geography, and/or cultural and linguistic factors.
- There was greater likelihood for ALS cases to be referred to ALS centers in SFBA versus LA.
  - Some smaller practices had cases to report, but did not participate. However, it is unclear if these unreported cases would have been unique.
- Characteristics from identified cases such as age of onset, slightly higher percentage of males, and higher proportion of white ALS cases are similar to existing literature.[4,6-8]
- A slightly lower than expected percentage of familial ALS was detected at 3.6% overall, compared to 5-10% frequently reported in the literature.[9]
- Race and ethnicity characteristics should be considered cautiously, especially for LA, which had higher rates of missing race and ethnicity data than SFBA.
- A review of death certificates for both areas and same time period identified 187 decedents with cause of death as ALS, but who were not among reported cases. It is unknown if these individuals were true ALS cases.
- Examining localized ALS incidence and demographics may help to reveal at-risk populations for additional studies.

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

CONCLUSION

This surveillance effort collected ALS data from two large metropolitan areas, with ethnically diverse populations. Data suggest that overall ALS incidence is comparable to current literature. However, more studies are needed to assess whether demographic patterns found in these regions represent true disease patterns. The incidence rate for LA should be interpreted with caution as fewer than expected cases were reported. The differences in the two regions with respect to the number of ALS cases collected may be explained by racial and ethnic differences or other factors like access to care, referral patterns, and geography. This project informs the larger national ALS surveillance effort, which is an important next step for better understanding the current epidemiology of ALS in the U.S. population.

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