Detroit 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 Detroit project.

METHODS

McKing partnered with the Detroit Department of Health and Wellness Promotion (DHWP) to conduct the project. All neurologists practicing in Wayne, Macomb, Oakland, and Washtenaw Counties, Michigan were asked if they diagnosed or provided care for ALS patients that resided in Wayne County. Emphasis was placed on neurologists specializing in the diagnosis/care of persons with ALS who practice at the four referral centers in the region 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. Crude incidence rates were calculated using the count of cases diagnosed in each year as the numerator and the corresponding U.S. Census population data[3] as the denominator. Crude average annual incidence rates were calculated by adding the incidence rates for the three years and then dividing by three. This project was approved by the Centers for Disease Control and Prevention Institutional Review Board.

RESULTS

- Twenty-eight percent (95/335) of neurologists indicated that they diagnosed and/or cared for ALS patients and seven reported cases. All major referral centers in the region participated.
- Using 2010 U.S. Census population data and estimates of incidence and prevalence, we expected to identify 146 unique cases in the project area.[3,4] A total of 172 case reports were received; 144 were unique cases, which is approximately 99% (144/146) of the expected cases.
- Eighty-seven percent (125/144) of cases were reported as "definite," "probable," or "probable-lab supported" according to the El Escorial criteria.[5] All 31 requested MRVFs were received and 84% (26/31) were classified as "definite," "probable," or "probable-lab supported," and 16% (5/31) were classified as "possible" by the consulting neurologist.
- Eighty-four percent of cases were 50 years of age or older at diagnosis, 58% were male, 76% were white, and 56% were not Hispanic or Latino (Table).
- Of the 113 cases for whom data were available, 50% had symptoms for 12 months or less before diagnosis. Ninety percent of the 113 cases were diagnosed within 44 months of having symptoms.
- Twenty-two percent (31/144) of cases had only federal payers [(Medicare, Medicaid, Veterans Affairs (VA)], 28% (41/144) had only non-federal payers (HMO, private insurance, self-pay, or other), and 50% (72/144) had both federal and non-federal payers.

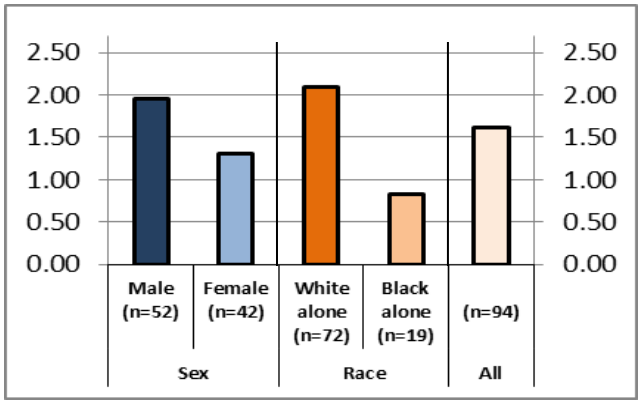
- There were 94 cases diagnosed in 2009-2011. The crude incidence rates for 2009, 2010, and 2011 ranged from 1.43 to 1.87 cases per 100,000 person-years. The crude incidence rates were not different by sex or race (Figure). The age-adjusted average annual incidence rate for the three year period was 1.61 cases per 100,000 person-years.

Table: Demographic Characteristics of All Reported ALS Cases in Wayne County, Michigan, n=144

Demographic Characteristic	n	%*
Age (years)		
Under 40	7	4.9
40 – 49	16	11.1
50 – 59	48	33.3
60 – 69	35	24.3
70 – 79	26	18.1
80 or older	12	8.3
Sex		
Male	84	58.3
Female	60	41.7
Race**		
White alone	109	75.7
Black/African American alone	32	22.2
Asian alone	1	0.7
Unknown	2	1.4
Ethnicity		
Hispanic	2	1.4
Not Hispanic or Latino	81	56.3
Unknown	61	42.4

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

Figure: Stratified Crude Average Annual Incidence Rates for ALS Cases Diagnosed in 2009-2011 in Wayne County, Michigan, n=94



The shaded bars represent the subgroup rates.
Incidence rates are per 100,000 person-years.

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

DISCUSSION

- All ALS referral centers in the region participated which was crucial to the success of the project.
- A small percentage of neurologists diagnosed or cared for patients with ALS. 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.
- Seventeen 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.
- We found higher, but not statistically significant, crude average annual incidence rates among ALS cases that were older, male, and white, which is consistent with published literature.[4,6,7]
- Examining localized ALS incidence and demographics may help to reveal at-risk populations for additional studies.

REFERENCES

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