Baltimore 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 Baltimore project.

METHODS

All neurologists practicing in the City of Baltimore, Baltimore County, and Howard County, Maryland were asked if they diagnosed or provided care for ALS patients that resided in the City of Baltimore, Baltimore County, and Howard County. Emphasis was placed on neurologists specializing in the diagnosis/care of persons with ALS who practice at 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.
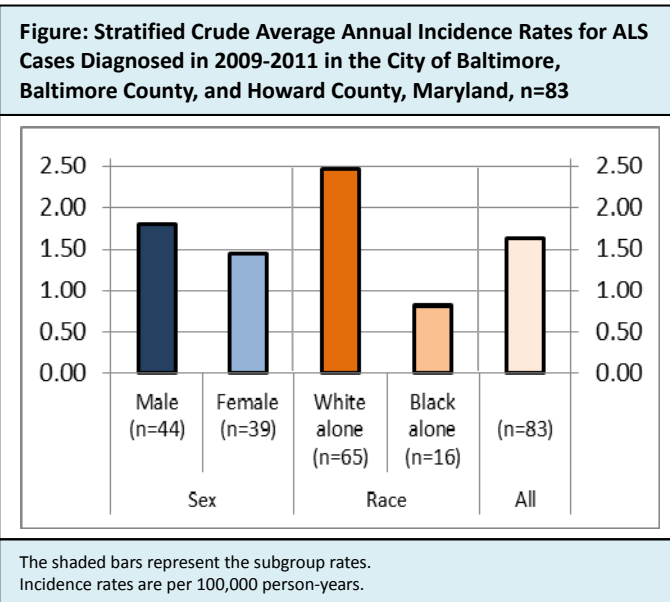
RESULTS

► Ten percent (33/318) of neurologists indicated that they diagnosed and/or cared for ALS patients and 24% (8/33) of those neurologists 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 137 unique cases in the project area.[3,4] A total of 144 case reports were received; 134 were unique cases, which is approximately 98% (134/137) of the expected cases.

► Ninety-one percent (102/134) of cases were reported as "definite," "probable," or "probable-lab supported" according to the El Escorial criteria.[5] Eighty-eight percent (15/17) of the requested MRVFs were received; 87% (13/15) were classified as "definite," "probable," or "probable-lab supported," and 13% (2/15) were classified as "possible" by the consulting neurologist.

► Eighty-three percent of cases were 50 years of age or older at diagnosis, 55% were male, 74% were white, and 97% were not Hispanic or Latino (Table).

► Of the 126 cases for whom data were available, 50% had symptoms for 12 months or less before diagnosis. Ninety percent of the 126 cases were diagnosed within 36 months of having symptoms.

► Sixteen percent (22/134) of cases had only federal payers [Medicare, Medicaid, Veterans Affairs (VA)], 24% (32/134) had only non-federal payers (HMO, private insurance, self-pay, or other), and 60% (80/134) had both federal and non-federal payers.

► There were 83 cases diagnosed in 2009-2011. The crude incidence rates for 2009, 2010, and 2011 ranged from 1.58 to 1.63 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 1.48 cases per 100,000 person-years.

Table: Demographic Characteristics of All Reported ALS Cases in the City of Baltimore, Baltimore County, and Howard County, Maryland, n=134		
Demographic Characteristic	n	%
Age (years)		
Under 40	9	6.7
40 – 49	12	9.0
50 – 59	29	21.6
60 – 69	43	32.1
70 – 79	23	17.2
80 or older	16	11.9
Unknown	2	1.5
Sex		
Male	73	54.5
Female	61	45.5
Race		
White alone	99	73.9
Black/African American alone	29	21.6
Asian alone*	2	1.5
Unknown	4	3.0
Ethnicity		
Hispanic	0	n/a
Not Hispanic or Latino	130	97.0
Unknown	4	3.0
*Those with multiple races are listed here.		

Figure: Stratified Crude Average Annual Incidence Rates for ALS Cases Diagnosed in 2009-2011 in the City of Baltimore, Baltimore County, and Howard County, Maryland, n=83

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 and submitted the vast majority of case reports.

► A small percentage of neurologists diagnosed or cared for patients with ALS. Some non-referral center practices in the region participated. The Baltimore VA Medical Center had cases to report, but only reported decedents. 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.

► Twenty-three 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 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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