
# New Jersey State-based 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 New Jersey project.

## METHODS

McKing partnered with the New Jersey Department of Health (NJDOH) to conduct the project. All neurologists practicing in the states of New Jersey and Delaware, two counties in New York (NY), and Philadelphia County, Pennsylvania (PA), as well as neurologists specializing in the diagnosis/care of persons with ALS practicing at ALS referral centers in Allentown, PA; New York City, NY; and Long Island, NY that typically see more than 50 patients per year, were asked if they diagnosed or provided care for ALS patients. 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 (IRB) and determined to be public health practice not requiring review by the NJDOH IRB.

## RESULTS

- Twenty-five percent (168/679) of neurologists indicated that they diagnosed and/or cared for ALS patients and 90% (152/168) 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 703 unique cases in the project area.[3,4] A total of 965 case reports were received; 764 were unique cases, which is approximately 109% (764/703) of the expected cases.
- Seventy-nine percent (600/764) of cases were reported as "definite," "probable," or "probable-lab supported" according to the El Escorial criteria.[5] Ninety-six percent (116/121) of the requested MRVFs were received; 82% (95/116) were classified as "definite," "probable," or "probable-lab supported," and 18% (21/116) were classified as "possible" by the consulting neurologist.
- Eighty-four percent of cases were 50 years of age or older at diagnosis, 55% were male, 83% were white, and 90% were not Hispanic or Latino (Table).
- Of the 752 cases for whom data were available, 50% had symptoms for 12 months or less before diagnosis. Ninety percent of the 752 cases were diagnosed within 36 months of having symptoms.
- Twenty-nine percent (225/764) of cases had only federal payers [Medicare, Medicaid, Veterans Affairs (VA)], 33% (255/764) had only non-federal payers (HMO, private insurance, self-pay, or other), and 37% (284/764) had both federal and non-federal payers.

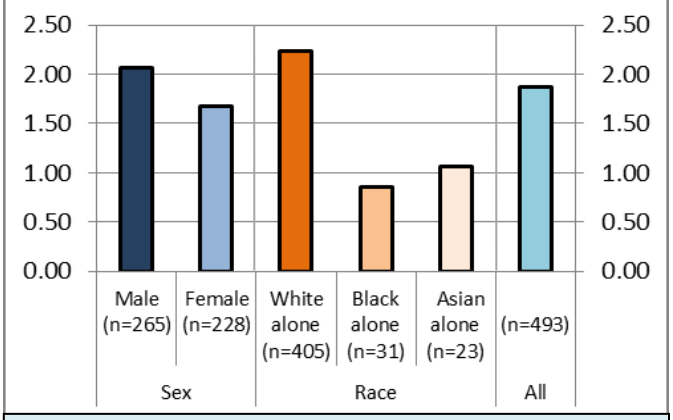
- There were 493 cases diagnosed in 2009-2011. The crude incidence rates for 2009, 2010, and 2011 ranged from 1.77 to 1.98 cases per 100,000 person-years. The crude average annual incidence rates between whites and Blacks/African Americans and between whites and Asians were significantly different (Figure). The age-adjusted average annual incidence rate for the three-year period was 1.67 cases per 100,000 person-years.

**Table: Demographic Characteristics of All Reported ALS Cases in New Jersey, n=764**

| Demographic Characteristic | n | %* |
|---|---|---|
| **Age (years)** | | |
| Under 40 | 35 | 4.6 |
| 40 – 49 | 87 | 11.4 |
| 50 – 59 | 184 | 24.1 |
| 60 – 69 | 210 | 27.5 |
| 70 – 79 | 172 | 22.5 |
| 80 or older | 71 | 9.3 |
| Unknown | 5 | 0.7 |
| **Sex** | | |
| Male | 421 | 55.1 |
| Female | 343 | 44.9 |
| **Race** | | |
| White alone | 636 | 83.2 |
| Black/African American alone | 59 | 7.7 |
| Asian alone | 33 | 4.3 |
| Other** | 7 | 0.9 |
| Unknown | 29 | 3.8 |
| **Ethnicity** | | |
| Hispanic | 44 | 5.8 |
| Not Hispanic or Latino | 687 | 89.9 |
| Unknown | 33 | 4.3 |

*May not add up to 100% due to rounding.
**Those with multiple races are listed here.

**Figure: Stratified Crude Average Annual Incidence Rates for ALS Cases Diagnosed in 2009-2011 in New Jersey, n=493**




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

- One-quarter of neurologists diagnosed or cared for patients with ALS during the reporting period and 90% of them reported cases.
- All ALS referral centers in the region participated and submitted the majority of case reports.
- Some non-referral center practices in the region participated. Some non-referral center practices and the VA hospitals and clinics declined to participate. However, it is unclear if providers at these practices would have reported unique ALS cases.
- Many 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 crude annual incidence rates among ALS cases that were older, male, and white, which is consistent with published literature.[4,6,7] The difference in crude rates between whites and Blacks/African Americans and between whites and Asians was statistically significantly different.
- Examining localized ALS incidence and demographics may help to reveal at-risk populations for additional studies.

## REFERENCES


1. National Amyotrophic Lateral Sclerosis (ALS) Registry. Centers for Disease Control and Prevention/Agency for Toxic Substances and Disease Registry Web site. http://wwwn.cdc.gov/als. Updated January 17, 3013. Accessed May 15, 2014.
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