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Atlanta metropolitan area amyotrophic lateral sclerosis (ALS) surveillance: incidence and prevalence 2009–2011 and survival characteristics through 2015

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Abstract

Amyotrophic lateral sclerosis (ALS) is a fatal, rare, and hard to diagnose neurological disease with unknown etiology.

Objective: To understand the incidence, prevalence, and survival characteristics of ALS cases in the Atlanta metropolitan area.

Methods: Neurologists in Clayton, Cobb, DeKalb, Fulton, and Gwinnett counties provided case reports for ALS patients under their care from 1 January 2009 to 31 December 2011. Incidence and prevalence rates were calculated for 2009, 2010, and 2011 by sex, race, and ethnicity. Using data from the National Death Index, survival time was calculated for age, sex, race, ethnicity, and El Escorial criteria.

Results: There were 281 unique ALS cases reported, which is approximately 104% of the expected cases. The majority of the 281 cases were white, non-Hispanic, male, and in the 50–59 age category. The overall average incidence rate for 2009 to 2011 was 1.54 per 100,000 person-years, with higher annual incidence rates for whites, males, and non-Hispanics. The prevalence rates for 2009, 2010, and 2011 were 5.05, 5.44, and 5.56 per 100,000, respectively. Median survival time was highest for the 18–39 age group, Asians, non-Hispanics, and males. Additionally, the log-rank tests for homogeneity across strata indicate a statistical significance between strata for the age category for survival time.

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Disclaimer

The findings and conclusions in this report are those of the authors and do not necessarily represent the official positions of ATSDR, CDC, and/or the HHS.

Declaration of interest

The authors declare no conflicts of interest. The conclusions of this article are those of the authors and do not necessarily represent the views of ATSDR, the Centers for Disease Control and Prevention (CDC), or the U.S. Department of Health and Human Services (HHS).

Conclusion: The findings for Atlanta are similar to other population-based studies in the United States. Although the Atlanta metropolitan area was selected to over-represent the minority population, the strongest predictor of survival time was age at diagnosis.

Keywords

Amyotrophic lateral sclerosis (ALS); surveillance; minority populations; incidence; prevalence; survival characteristics; Atlanta; National ALS Registry

Introduction

Amyotrophic lateral sclerosis (ALS) is a progressive and fatal neurological disease affecting both the upper and lower motor neurons. Familial forms of ALS account for 5–10% of cases (1,2). The etiology of most sporadic ALS cases is unknown, and previous etiologic studies have evaluated environmental and occupational exposures, physical activity, trauma, genetics, as well as other potential risk factors (3-5).

ALS prevalence is estimated to be 4–6 cases/100,000 population globally, with Japan reporting one of the highest crude prevalence rates of 9.9 per 100,000 population (6-8). Recent data from the National ALS Registry estimated the prevalence of ALS in the United States (US) to be 5.2 per 100,000 population (9). Worldwide prevalence and incidence rates may vary widely due to differences in population age and geography, including genetic predisposition, and environmental factors (7). In North America, the annual incidence rate is estimated to be 1.6 cases per 100,000 person-years (7,10). Those with ALS in the United States are more likely to be older, male, white, and non-Hispanic (6,9,11,12). Median survival time from diagnosis has been reported to be 21–26 months depending on the population studied (13-15). The strongest predictor of survival time has consistently been age at diagnosis, with older ages having shorter survival time (13-16).

To learn more about ALS in the United States, the federal Agency for Toxic Substances and Disease Registry (ATSDR) maintains the National ALS Registry (Registry) (9,17). Because ALS, like most noncommunicable diseases, is a non-notifiable condition in the United States, the Registry identifies ALS cases using national administrative databases and by patient self-enrollment through a web portal (9,17,18). ATSDR's Registry also funded the State and Metropolitan Area ALS Surveillance Project, which consisted of three state and eight metropolitan areas in the US, including Atlanta. The purpose of this project, described in detail elsewhere (19), was to gather reliable and timely data on the demographic characteristics of ALS cases in smaller catchment areas below the national level and to assist ATSDR in evaluating the completeness of the Registry.

The objectives of this paper are to calculate the incidence and prevalence of ALS in the Atlanta metropolitan area and describe the demographic and survival characteristics of cases to contribute to the epidemiologic knowledge of ALS in a diverse population.

Methods

Study population

The data presented here were collected as part of the overall State and Metropolitan Area ALS Surveillance Project. Catchment areas were selected to over-represent racial and ethnic minorities. The Atlanta metropolitan catchment area included the five largest counties, Cobb, Clayton, DeKalb, Fulton, and Gwinnett with a total population of 3,365,297. All neurologists in the catchment area were contacted to determine if they diagnosed and/or cared for ALS patients during the eligibility period. If they did, a surveillance coordinator explained the surveillance project, identified a contact, and requested that each eligible person with ALS be reported. A person with ALS was eligible to be reported if diagnosed and/or cared for from 1 January 2009 through 31 December 2011 and was a resident of one of the project's catchment areas.

All neurologists who diagnosed or cared for ALS patients were requested to complete a one-page Case Reporting Form (CRF). Additional focus was given to neurologists who typically see more than 50 ALS patients per year. The CFR included patient demographic questions such as sex, date of birth, race, ethnicity, date of diagnosis, El Escorial criteria, date of onset of symptoms, family history, provider details, and payer information. Race was categorized using the US Census defined race categories. Race and ethnicity were self-reported variables abstracted from the patient's medical records. The El Escorial diagnosis criteria was used for case definition, where patients were categorized as definite, probable, probable (lab supported), possible, or not classifiable ALS (20). ALS diagnosis was confirmed through a medical record verification form (MRVF) and an electromyogram (EMG) report. No patients were directly contacted as a part of the overall State and Metropolitan Area ALS Surveillance Project. The age of diagnosis was not reported for the metropolitan areas but calculated using the date of birth and the date of ALS diagnosis. For the analyses, age was categorized into six groups 18–39, 40–49, 50–50, 60–69, 70–79, and greater than 80 years of age. All cases identified in the Atlanta metropolitan area were submitted to the National Death Index (NDI) and searched for deaths occurring in years 2009–2015 to learn vital status and identify the date of death for deceased cases. A detailed description of the methodology used for case ascertainment can be found in the State and Metropolitan Area ALS Surveillance Project paper by Wagner et al (19).

Incidence

Incidence rates were calculated for each year using the count of cases diagnosed in each year as the numerator and the corresponding county 2010 US Census population data as the denominator. Only cases diagnosed from 2009 to 2011 were included, excluding those cases diagnosed prior to 2009. Crude average annual incidence rates were calculated by adding the incidence rates for the three years and then dividing by three. Furthermore, cases were stratified by sex, race, and ethnicity and strata-specific incidence rates were calculated for 2009, 2010, and 2011. An average annual incidence rate (2009–2011) was calculated for sex, race, and ethnicity. All incidence cases are calculated and listed as cases per 100,000 person-years.

Prevalence

Prevalence was calculated for 2009, 2010, and 2011 using case data collected during the State and Metropolitan Area Surveillance Project. Cases were submitted to the NDI to obtain information on vital status and, if deceased, obtain date of death. Using the data received from the NDI, those cases who were deceased were excluded from the appropriate yearly prevalence calculation. Six cases missing date of diagnosis were excluded from the prevalence analysis. Unknown, other race, and unknown ethnicity were excluded from the prevalence analysis. The 2010 US Census population for the five counties was used for the denominator and standardized to 100,000 population.

Survival time

A survival analysis was conducted using data from the cases in the Atlanta metropolitan area and date of death obtained from the NDI 2015. In this analysis, survival time was calculated from the month and year of diagnosis to the month and year of death and reported in months of survival. Cases identified as unknown or other race were excluded from the analysis. The ethnicity variable included Hispanic and non-Hispanic.

Using the LIFETEST procedure in SAS version 9.3, median and mean survival times were generated. The variables of age group, sex, race, ethnicity, and El Escorial diagnosis criteria were analyzed. Because neurologists specializing in the treatment of ALS agree that patients with El Escorial diagnosis criteria definite, probable, and probable (laboratory-supported) have ALS, we combined them into one category and possible and not classifiable into the second category. The median survival time was reported (in months), along with the confidence intervals for the function. Using the log-rank test, survival time was assessed across strata and a p-value less than 0.05 was considered statistically significant. Data were analyzed for descriptive and survival analysis using Microsoft Excel and SAS software version 9.3 (21,22). This project was approved by the Centers for Disease Control and Prevention Institutional Review Board.

Results

Case demographics

Of the Atlanta catchment area, the 2010 Census population was 45.9% white and 39.1% black, with 87.5% non-Hispanic and 12.5% Hispanic. The 2010 US population in comparison was 72.4% white and 12.6% black, with 83.7% non-Hispanic and 16.3% Hispanic (23). Out of a total of 226 neurologists, only 30 neurologists in the catchment area diagnosed or cared for ALS patients and 87.0% (26 neurologists) reported cases. A total of 292 case reports were received from the participating neurologists representing 281 unique cases that were used in this analysis, which was 104% of expected cases. In order to evaluate the progress of the numerous state and metropolitan areas, we estimated the number of expected cases by using the 2010 Census data for the project area and estimates of incidence and prevalence.

Two-hundred twenty-nine of the 281 (81.5%) cases were reported as being definite, probable, or probable lab-supported cases on the El Escorial criteria. The remaining 52 cases

were defined as possible or not classifiable cases. Overall, 45.6% of the reported Atlanta patients were over the age of 60 years compared with a higher national average of 62.7% of cases (24). Comparing the sex distribution of ALS cases, the Atlanta cohort had a 1.6 male to female ratio compared with a similar 1.5 male to female ratio for the national data. The majority of Atlanta patients were white (69.4%), followed by black (23.1%), and Asian (2.8%). Eighty-nine percent of ALS cases were non-Hispanic, and the majority classified as definite on the El Escorial criteria (58.4%). The reported Atlanta cases were most likely to be between the ages of 50–79, male, white, and non-Hispanic (Table 1). Familial history of ALS was reported for 3.9% of the cases.

Incidence

Of the 281 unique cases reported, 155 (55.2%) cases were diagnosed between 1 January 2009 and 31 December 2011 in the Atlanta area. Using the 2010 US Census population, the incidence rates for each year and an overall annual incidence rate was calculated. The overall crude incidence rates for 2009, 2010, and 2011 were 1.49, 1.66, and 1.46 per 100,000 person-years, respectively. The average incidence rate for the three-year period was 1.54 per 100,000 person-years (Table 2).

Additionally, incidence rates were analyzed by race, sex, and ethnicity. Males had a higher overall annual incidence rate of ALS (1.88 cases per 100,000 person-years) compared with women (1.22). Additionally, the average incidence rate for whites was 2.46 compared with 0.94 for blacks and 0.62 for Asians. The average incidence rate for non-Hispanic was 1.66 cases per 100,000 population compared with 0.47 for the Hispanic cases.

Prevalence

In 2009, a total of 170 prevalent ALS cases were reported by neurologists in the Atlanta area. From 2009 to 2011, the number of prevalent cases of ALS increased from 170 to 187 cases. Those cases identified by the neurologists in 2009 also counted in 2010, unless the case was confirmed deceased by the NDI. The annual prevalence for 2009 was 5.05 per 100,000 population, with males having a higher prevalence (6.60) than women (3.58) (Table 3). The prevalence rate for 2010 increased to 5.44 per 100,000 population, followed by 5.56 for 2011. Across 2009 to 2011, the strata-specific prevalence rates tend to be highest for white, non-Hispanic, and males (Table 3).

Survival characteristics

The final analysis consisted of calculating survival time for the Atlanta cohort of cases. As of 31 December 2015, 204 (72.6%) of the reported cases were deceased. The overall mean survival time from the time of diagnosis to the time of death was 34.6 months and the median survival time was 23 months (95% CI 21, 28) (Table 4). Survival time varied among the age groups, with survival time decreasing with increasing age. The highest survival time (54.5 months) was for those 18–39 years of age. The survival time was the lowest for those ≥80 years of age (10 months 95% CI 3, 21). Males had a higher median survival time of 26 months (95% CI 21, 34) compared with females with a survival time of 21 months (95% CI 14, 25). The survival time for Hispanics was much longer compared with non-Hispanics with 48 months (95% CI 37, 55) and 27 months (95% CI 21, 37), respectively.

Finally, those cases reported as definite, probable, and probable (laboratory supported) on the El Escorial criteria had a slightly higher median survival time of 24 months (95% CI 21, 29) compared with 21 months (95% CI 10, 29) for those in the possible and not classifiable diagnosis group. The log-rank tests for homogeneity across strata indicate a statistical significance between strata for the age category with a p-value of $p \leq 0.0001$ (Table 4).

Discussion

This report is the first to provide incidence and prevalence estimates specific to the Atlanta metropolitan area. Although every attempt was made to identify ALS cases in the area, it is possible that a small percentage of ALS patients residing in the Atlanta metropolitan area were not included. However, because the ALS specialty clinics in Georgia both reported their eligible cases which comprised of 95% of the unique cases and the number of neurologists who would care for an ALS patient who did not report was small, 4, the chance that underreporting bias occurred or a large number of cases were missed is likely to be small.

The demographic characteristics of the Atlanta cohort were similar to previously published literature (6,7,14). The annual incidence rates for Atlanta (2009–2011) ranged from 1.49 to 1.66, with an average annual incidence rate of 1.54 per 100,000 per-years. This incidence is similar to the worldwide estimates of 1.6–2.5 per 100,000 population (7,8,25,26). The annual average incidence rate for the Atlanta metropolitan cohort was higher for males than females, which is similar to previously published literature (7,25,26). Compared internationally, the European population had a crude incidence rate of 2.16 per 100,000 person-years (95% CI 2.0, 2.3), with men also having a higher incidence rate compared with women (25). In addition, those who reported their race as white had a significantly higher incidence rate compared with black and Asian. A lower incidence for blacks, Asians, and non-Hispanic ALS patients is consistent with other published data analyzing the difference in incidence rates among ethnicities (12,27). This reported lower incidence in minorities and Atlanta's higher percentage of a minority population may explain the slightly lower average annual incidence rate reported in this analysis.

To our knowledge, this project was the first to analyze ALS prevalence rates in the Atlanta metropolitan area. In addition to incidence rates per year, the prevalence rates for the Atlanta metropolitan ALS cases for 2009, 2010, and 2011 were calculated. The overall prevalence in 2011 (5.56 per 100,000) was higher than the national prevalence rate of 4.3 per 100,000 (24). Additionally, a global analysis conducted of motor neuron diseases from 1990 to 2016 found a worldwide prevalence rate of 4.5 (4.1–5.0) per 100,000 population, which is lower than the prevalence rates determined in the Atlanta cohort (28). Similar to other ALS literature, the prevalence of white cases in Atlanta (2009–2011) is significantly higher compared with the other races, with a significant difference in the number of Hispanic ALS patients seen by neurologists compared with non-Hispanic patients (7,27). When stratifying the prevalence by race, sex, and ethnicity, the prevalence for males in the Atlanta cohort is significantly higher compared with the prevalence for females, which is similar to existing literature (7,9,10).

The survival analysis indicates median survival time for the Atlanta cohort to be 23 months from the time of diagnosis, which is similar to other cohort studies reporting average survival times of 21–26 months (13,14). A study conducted on ancestral origin of ALS cases reveals a median survival time from ALS onset to be highest for those from Central Asia (48 months) and the lowest survival for cases from Northern Europe (25 months) (29). Although the number of Asians in the Atlanta area were low and not statistically significant, like other papers, Asians had the highest survival time compared with blacks or whites. Additionally, other studies have concluded lower mortality rates for mixed origin populations compared with the white population (30). The survival time for men in the Atlanta cohort was higher than woman, which is similar to findings from other studies assessing survival time characteristics (16). However, the age at diagnosis was the only statistically significant factor for survival time, with survival time decreasing as the age of diagnosis increases. Other studies have also concluded that increasing age is a strong predictor of survival time (13,14).

A small number of cases in the unknown and other race category, as well as the unknown ethnicity were excluded from the analysis. The Atlanta metropolitan area was selected to over represent racial and ethnic minority populations. While the US population consists of 72.4% white, 12.6% black, and 4.8% Asian, the Atlanta population has a significantly higher percentage of blacks (39.1%) and Asians (6.4%) (23). When comparing the percentage of the Hispanic population, there is a slightly lower percentage of Hispanics in Atlanta (12.5%) compared with the United States (16.3%). Because Hispanics account for 12.5% of the population in the Atlanta counties, with an emphasis towards a younger Hispanic population, the low percent of Hispanic ALS cases is not unexpected. Looking at the percentage of non-whites in the United States and Atlanta counties, if race and ethnicity were predictive of survival time, the Atlanta cohort had a sufficient sample of non-whites to evaluate if race was a predictor of survival time. However, with an adequate sample size, there was still no statistical significance for survival time by race or ethnicity. The results and findings from the Atlanta metropolitan area are consistent with other similar metropolitan areas participating in the State and Metropolitan Area ALS Surveillance project (11,28,31,32). As in the Atlanta cohort, age was the strongest predictor of survival in other similar metropolitan areas (13-16).

We found that those cases reported as definite, probable, and probable (lab supported) on the El Escorial criteria had a slightly higher median survival time compared with those in the possible and not classifiable diagnosis group. Studies have shown that individuals who regularly are seen at an ALS specialty clinic have longer survival (33,34). Although we do not know who regularly attended a specialty clinic, we can hypothesize that those with a less definitive diagnosis are less likely to attend such a clinic which could contribute to their decreased survival time.

Conclusion

Conducting surveillance for ALS in the United States can be a challenge because ALS is a non-notifiable disease and systems developed to identify cases may not be complete. However, this project identified 104% of the expected cases in the Atlanta metropolitan area, suggesting that there was minimal under-ascertainment. These analyses are the first to

examine the incidence, prevalence, and survival time for the Atlanta cohort from the State and Metropolitan Area ALS Surveillance Project. Although the Atlanta metropolitan area was selected to over-represent racial and ethnic minorities, the results are similar to other national findings in the United States, with an overall annual incidence rate of 1.54 per 100,000 person-years and prevalence rates ranging from 5.05 to 5.56 per 100,000 population for 2009 to 2011. While there were some differences when assessing survival time by race, sex, ethnicity, and El Escorial criteria, the strongest predictor was age at diagnosis. The findings from these analyses contribute the knowledge of ALS incidence, prevalence, and survival time in a diverse US population.

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Table 1.

Demographic characteristics of reported ALS cases in the Atlanta* metropolitan area, 1 January 2009 to 31 December 2011 (n=281).

		Count of Cases (% of cases)	
Demographics	**Categories**	**#**	**%**
Age Groups (Years)	18–39	22	7.8
	40–49	50	17.8
	50–59	75	26.7
	60–69	71	25.3
	70–79	48	17.1
	≥80	9	3.2
	Unknown	6	2.1
Sex	Male	172	61.2
	Female	109	38.8
Race	White	195	69.4
	Black	65	23.1
	Asian	8	2.8
	Other	1	0.4
	Unknown	12	4.3
Ethnicity	Hispanic	9	3.2
	Non-Hispanic	250	89.0
	Unknown	22	7.8
El Escorial Diagnosis Criteria	Definite	164	58.4
	Probable	45	16.0
	Probable (laboratory supported)	20	7.1
	Possible	45	16.0
	Unclassified	7	2.5
Family History of ALS	Yes	11	3.9
	No	259	92.2
	Unknown	11	3.9
Total		281	100

*Atlanta metropolitan area includes Cobb, Clayton, DeKalb, Fulton, and Gwinnett counties.

Table 2.

Annual incidence rates for ALS cases diagnosed in a 3-year period in the Atlanta metropolitan area, 2009–2011 (n=155).

	2009		2010		2011		2009–2011
Characteristic	**Cases**	**Incidence**	**Cases**	**Incidence**	**Cases**	**Incidence**	**Average Incidence per 100,000 person years**
Sex							
Male	31	1.90	32	1.96	29	1.77	1.88
Female	19	1.10	24	1.39	20	1.16	1.21
Race *							
White	36	2.33	42	2.72	36	2.33	2.46
Black	12	0.91	13	0.99	12	0.91	0.94
Asian	2	0.93	1	0.46	1	0.46	0.62
Ethnicity **							
Hispanic	0	0.00	2	0.47	4	0.95	0.47
Non-Hispanic	50	1.70	52	1.77	45	1.53	1.66
Total	50	1.49	56	1.66	49	1.46	1.54

* Excludes unknown and other race (n=13).

** Excludes unknown ethnicity (n=22).

Table 3.

Prevalence rates for 2009, 2010, and 2011 for the Atlanta metropolitan area.

		2009		2010		2011	
Categories	**US 2010 Census Population**	**Cases**	**Prevalence**	**Cases**	**Prevalence**	**Cases**	**Prevalence**
Sex							
Male	1,635,376	108	6.60	116	7.09	118	7.22
Female	1,729,921	62	3.58	67	3.87	69	3.99
Race							
White	1,546,399	115	7.44	130	8.41	137	8.86
Black	1,314,721	39	2.97	40	3.04	40	3.04
Asian	215,881	6	2.78	6	2.78	5	2.32
Ethnicity							
Hispanic	422,202	2	0.47	3	0.71	6	1.42
Non-Hispanic	2,943,095	150	5.10	167	5.67	170	5.78
Total *	3,365,297	170	5.05	183	5.44	187	5.56

* Includes those with unknown, other race, and unknown ethnicity.

Table 4.

Median survival time by demographic and diagnosis characteristics for the Atlanta metropolitan area (*n*=204).

Categories	Number of Cases	Median Survival Time (95% CI*) in Months	Mean Survival Time (SD)	Log-rank test of equality
Age Groups				
18–39	12	54.5 (19, 86)	72.5 (21.8)	$p \leq 0.0001$
40–49	31	38.0 (23, 49)	42.1 (4.73)	–
50–59	51	28.0 (18, 37)	36.7 (5.00)	–
60–69	58	24.0 (18, 31)	32.8 (4.03)	–
70–79	43	16.0 (11, 22)	23.2 (3.87)	–
≥80	9	10.0 (3, 21)	12.2 (2.24)	–
Sex				
Male	124	26.0 (21, 34)	37.7 (3.16)	$p=0.0874$
Female	80	21.0 (14, 25)	29.8 (4.05)	–
Race**				
Black	49	21.0 (14, 34)	38.7 (3.43)	$p=0.3771$
White	142	23.0 (21, 29)	35.1 (3.26)	–
Asian	6	38.0 (18, 124)	50.3 (16.9)	–
Ethnicity***				
Hispanic	4	48.0 (37, 55)	47.0 (3.74)	$p=0.4215$
Non-Hispanic	185	23.0 (20, 26)	33.2 (2.64)	–
El Escorial Diagnosis Criteria – Groups				
Definite, Probable and Probable (laboratory-supported)	176	24.0 (21, 29)	35.6 (2.80)	$p=0.3301$
Possible and Not classifiable	28	21.0 (10, 29)	28.3 (4.51)	–
Total	204	23.0 (21, 28)	34.6 (2.50)	–

* Confidence interval (CI).

** Excludes unknown and other race (*n*=7).

*** Excludes unknown ethnicity (*n*=15).