Findings of the New Jersey Amyotrophic Lateral Sclerosis Surveillance Project, 2009-2011

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Background - ALS

- Amyotrophic Lateral Sclerosis (ALS or Lou Gehrig's disease) is a rare, progressive, incurable neurological disease
- Complex etiology with no definitive cause; small proportion of cases (5-10%) associated with inheritance (termed familial ALS)
- Difficult to diagnose
  - Definitive clinical lab test does not exist
  - Diagnosed based on the use of the El Escorial criteria, which considers a combination of symptoms and electromyogram (EMG) reports to reach a determination of possible, probable, probable-lab supported, or definite ALS

Background - Epidemiology

Crude incidence	1.6 (range 0.7-2.5) cases per 100,000 person-years[1]
Crude prevalence	4.0 cases per 100,000 persons[1]
Mean age at disease onset	mid-60s; most people who develop the disease are between the ages of 55 & 75[1-5]
Demographic distribution	Rates higher among those who are older, male, white, and non-Hispanic[1,5-7]
Median time from symptom onset to diagnosis	9-11 months[2]
Median survival from diagnosis	16.5-23 months (<10% survive 5 years or longer)[2]

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2. Mitsumoto H, Chad DA, Pioro EP. (1998). Amyotrophic Lateral Sclerosis. Philadelphia, PA: F.A. Davis Company.
3. Eisen A. Amyotrophic Lateral Sclerosis is a multifactorial disease. Muscle Nerve. 1995;18:741-52
4. Chio A. Risk factors in the early diagnosis of ALS: European epidemiological studies. Amytroph Lateral Scler Other Motor Neuron Disord. 2000;1Suppl1:S13-8
5. Mehta P, Antao V, Kaye W, et al. Prevalence of amyotrophic lateral sclerosis--United States, 2010-2011. MMWR Morb Morta Wkly Rep 2014;63(No. SS-7):1-13
6. Cronin, S, Hardiman O, Traynor BJ. Ethnic variation in the incidence of ALS: a systematic review. Neurology. 2007;68:1002-1007
7. Gundogdu B, Al-Lahham T, Kadlubar F, Spencer H, Rudnicki SA. Racial differences in motor neuron disease. Amyotroph Lateral Scler Frontotemportal Degener. 2014;15:114-118

Background - Registry

- The federal Agency for Toxic Substances and Disease Registry (ATSDR) maintains the congressionally-mandated National ALS Registry (Registry)
- Because ALS is a non-notifiable disease in the US, the Registry identifies cases using national administrative datasets and patient self-registration through a web portal
- This non-traditional case ascertainment required validation against conventional surveillance techniques

Background - Registry

- ATSDR established three state and eight metropolitan-area ALS Surveillance Projects
- The objectives of the projects were to calculate the incidence and prevalence of ALS, and to gain a better understanding of demographic characteristics of ALS
- This presentation describes the New Jersey (NJ) project

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Methods

- All neurologists in NJ, DE, some counties in downstate NY, New York City, NY, Philadelphia, PA, and Allentown, PA were contacted to determine if they diagnosed or provided care to ALS patients and were asked to report eligible cases
- Eligible cases were under the doctor's care between 1/1/2009-12/31/2011, could be assigned to an El Escorial criteria classification level, and were residents of NJ
- A medical record verification form and a copy of an EMG report were requested for a sample of cases to evaluate the accuracy of the diagnosis

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Methods

- Multiple case reports for the same person were accepted from different practices and the dataset was de-duplicated prior to analysis
- Mortality records were queried for the period 2009-2013 using the ICD-10 G12.2 code for motor neuron disease and key term literals
- Compensation was offered for each submitted form
- No patients were contacted
- The project was approved by the CDC Institutional Review Board (IRB) and deemed public health surveillance by the NJ Department of Health (DOH) IRB

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Methods

- Stratified age-adjusted point prevalence rates on December 31, 2011 were calculated for cases with a known age at diagnosis and who were not known to be deceased on December 31, 2011
- Age-specific average annual incidence rates and stratified age-adjusted average annual incidence rates were calculated for cases with a known age at diagnosis and with a diagnosis between 2009 and 2011
- Incident cases were examined to determine if ALS clusters geographically

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Methods

- Geographic clustering of ALS incidence was examined using a spatial scan statistic at the census tract level
- Survival estimates by age, sex, race, and ethnicity were calculated using the Kaplan-Meier estimator for incident cases diagnosed between 2009 and 2011 and followed until death or December 31, 2013

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Results

Table 1: Recruitment and Participation of Neurologists

	New Jersey		Other[a]		Total	
	#	%	#	%	#	%
Total	429	100.0	250	100.0	679	100.0
Diagnosed or cared for ALS patients in reporting period	143	33.3	25	10.0	168	24.7
Reported cases	132	30.8	20	8.0	152	22.4
Did not report cases	11	2.6	5	2.0	16	2.4
Did not diagnose or care for ALS patients in reporting period	101	23.5	59	23.6	160	23.6
Does not diagnose or provide care for ALS patients	185	43.1	166	66.4	351	51.7
Other reporting physician	2	n/a	0	n/a	2	n/a

[a]All neurologists practicing in Delaware and some counties in New York, as well as ALS specialists in New York City, NY, Long Island, NY, Philadelphia, PA, and Allentown, PA; and reporting neurologists in Florida and Connecticut.

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Results

- 965 case reports were collected
- 21% (199) were cases reported more than one time and composite records were created for these cases
- Prevalence
  - 764 unique prevalent cases over the three year period were retained in the final data set
  - Age-adjusted point prevalence on December 31, 2011 was 4.40 cases per 100,000 persons
  - Rates were higher for males compared with females, Whites compared with Blacks and Asians, and non-Hispanics compared with Hispanics

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Results

- Incidence
  - 493 unique cases diagnosed in 2009-2011 were retained in the final data set
  - The average annual age-adjusted incidence rate was 1.67 cases per 100,000 persons
  - Rates were higher among males compared with females, Whites compared with Blacks and Asians, and non-Hispanics compared with Hispanics

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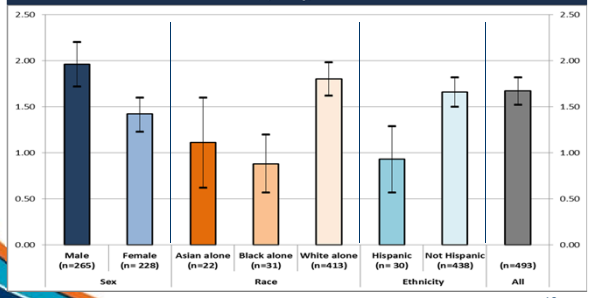
Results

Figure 1: Stratified Age-adjusted Average Annual Incidence Rates (per 100,000 Person-years) for ALS Cases Diagnosed in 2009, 2010, and 2011 in New Jersey, n=493



Results

Table 2: Age-specific Average Annual Incidence Rates for ALS Cases Diagnosed in 2009, 2010, and 2011 in New Jersey, n=493

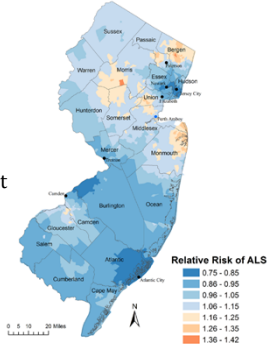
Age Category (in years)	Count of Cases	NJ 2010 Population[a]	Age-specific Rate Per 100,000 Person-years
Under 30	4	3,385,581	0.04
30 – 39	13	1,145,041	0.38
40 – 49	40	1,354,434	0.98
50 – 59	114	1,240,303	3.06
60 – 69	143	831,514	5.73
70 – 79	119	476,177	8.33
80 or older	60	358,844	5.57

[a]The total population in NJ in 2010 was 8,791,894.[16]

Results

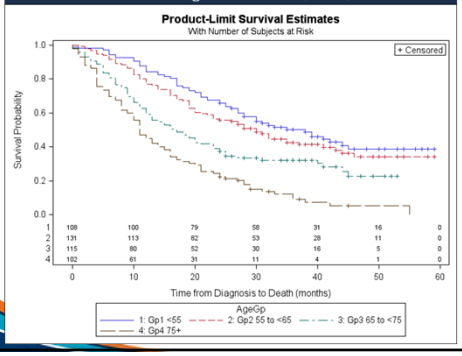
- No geographic clusters identified
- ALS incidence varied across and within counties; relative risk at the census tract level ranged from 0.75 to 1.42

Figure 2: Geographically Smoothed Relative Risk Map of Amyotrophic Lateral Sclerosis (ALS) in New Jersey, 2009-2011, n=493



Results

Figure 3: Kaplan-Meir Survival Curves for Incident ALS Cases in NJ (2009-2011), by Age Group at Diagnosis, with Vital Status Determined Through December 31, 2013, n=465



Results

- NJ DOH used these data to develop an ALS Project Summary
- Three published papers and several presentations at scientific meetings
- Incidence and prevalence data were synthesized to construct an ALS Indicator; after pilot-testing, the Indicator was uploaded for display on the NJ Environmental Public Health Tracking (EPHT) health data portal

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Results

Conclusions

- This project expands our scientific understanding of ALS occurrence, and provides the first state-wide ALS incidence, prevalence, and survival data by demographic groups
- Age-adjusted point prevalence, annual average age-adjusted incidence rates, and survival are consistent with previously published literature
- We found higher age-adjusted average annual incidence rates and age-adjusted point prevalence rates among ALS cases who were older, male, non-Hispanic and White, which is consistent with published literature

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Conclusions

- Men and women living in the wealthiest or highest-income areas of the state had a higher risk of ALS than those living in lower-income areas
- More research is needed to understand why census-track median income produces variability in risk of ALS
- Future studies should include individual and area-level measures of SES, behavior, occupational, and environmental risk factors to better understand disease etiology

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Conclusions

- Conducting time-limited surveillance for a non-reportable chronic condition was challenging and expensive
- With adequate planning, adhering to data collection methodologies, providing compensation, and executing quality assurance procedures, we successfully collected case reports for NJ residents for a non-reportable chronic disease from neurologists on a regional scale
- The project illustrates how EPHT partnerships can enhance chronic disease surveillance and help make chronic disease data available to the general public, health planners, and advocacy organizations

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Additional Information

- National ALS Registry: https://wwwn.cdc.gov/als/
- NJ Department of Health ALS Surveillance Project: http://www.nj.gov/health/als/index.shtml
- NJ State Health Assessment Data (NJSHAD): https://www26.state.nj.us/doh-shad/home/Welcome.html
- NJ Environmental Public Health Tracking (NJEPHT): http://www.state.nj.us/health/epht/

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Additional Readings

- Jordan H, Fagliano J, Lefkowitz D, Rechtman L, Kaye WE. Population-based surveillance of Amyotrophic Lateral Sclerosis in New Jersey, 2009-2011. Neuroepidemiology 2014;43(1):49-56. doi: 10.1159/000365850
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- Henry K, Fagliano J, Jordan H, Rechtman L, Kaye W. Geographic variation of Amyotrophic Lateral Sclerosis incidence in New Jersey, 2009-2011. American Journal of Epidemiology 2015;epub ahead of print. doi: 10.1093/aje/kwv095

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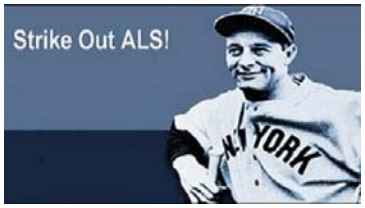
Thank You

Questions, comments, or feedback?



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