

Prevalence of Amyotrophic Lateral Sclerosis (ALS), United States, 2016

Frequently Asked Questions (FAQs)

1. Why is the National ALS Registry important?

The National ALS Registry does a lot more than just count ALS cases. It is a multi-faceted research platform. Some activities include:

- [funding research](#) to understand the causes and risk factors of ALS;
- supporting recruitment for [clinical trials and epidemiological studies](#) on behalf of pharmaceutical companies, ALS referral centers, and universities;
- collecting blood and tissue for the [National ALS Biorepository](#) to support and conduct research in the areas of genetics analyses, biomarker identification, and disease progression; and
- better defining the epidemiology of ALS in the United States, and determining how this information can be used to evaluate public health burden

2. What is the Prevalence of Amyotrophic Lateral Sclerosis (ALS) report?

The Prevalence of Amyotrophic Lateral Sclerosis (ALS) report provides information about the number of existing ALS cases for calendar year 2016. Along with other publications from the Registry, this report informs the public about epidemiological trends of ALS in the U.S.

3. What do the 2016 report's findings mean? In 2016, the National ALS Registry identified 16,424 adult persons with ALS. From this finding, we estimated that 5.2 of every 100,000 adults in the United States have ALS.

4. How does it compare to the 2015 report? The findings for 2016 were similar to the 2015 report. In 2015, slightly more cases (16,583) of ALS were identified than 2016 (16,424). This is a difference of 159 cases, which is not statistically significant. The prevalence of 5.2 adults with ALS for every 100,000 persons remained the same. Reports from 2010 through 2015 are available, please click [here](#).

5. Why are you just now reporting 2016 results when we are in 2021? The Registry receives information about ALS cases from the Centers for Medicare and Medicaid Services (CMS), the Veteran's Administration (VA), and patients who enroll directly with the Registry at www.cdc.gov/als, and there is typically a multi-year lag in the data before they are released to ATSDR. After the Registry receives the CMS and VA data, epidemiologists and statisticians clean and process the data through an algorithm to identify cases. Case data are then merged with Registry self-enrollment case data to form a final dataset. The final dataset is sent to the National Death Index to determine who has passed away so they can be removed as a case. From the time the Registry receives data from CMS and VA, it typically takes up to 24 months to remove duplicate information from the datasets and finalize the number of ALS cases. The pandemic also caused delays in receiving data. CDC/ATSDR is largely reliant upon other federal agencies to provide source data to ATSDR. The Registry moves as quickly as possible to make annual reports available once the data are compiled.

- 6. Is the Registry able to count everyone with ALS?** No. ALS is not a notifiable disease, that is, required to be reported to government authorities in the United States. CDC receives data from state health departments on most communicable diseases and a few non-communicable diseases (e.g., tuberculosis, HIV, or cancer), but not the number of ALS cases. Additionally, surveillance systems that cover a large area (e.g., state and national levels) are not typically able to capture 100% of all cases, even when the disease or condition is notifiable (e.g., cancer). However, we are taking steps to ensure that the Registry's data are as complete as possible.
- 7. Why is ALS not reportable to state health departments?** Each state sets their own criteria for what diseases are reportable to their health departments. Massachusetts is the only state where physicians must report an ALS diagnosis to state health officials. However, there are no states that currently notify CDC/ATSDR of ALS cases. CDC/ATSDR is working with the Massachusetts Department of Health to compare their data with National ALS Registry data, to improve the effectiveness of the Registry's data capture model.
- 8. How many cases is the Registry missing?** Because ALS is not a notifiable disease in the United States, it is challenging to estimate precisely how many cases are not included in the Registry. The Registry is currently utilizing different methods to identify how many cases are missing, such as capture-recapture and cross-referencing Registry data with other data sources (e.g., ALS Association, Muscular Dystrophy Association, Massachusetts ALS Registry). Capture-recapture is a statistical method that measures the overlap of cases from different data sources.
- 9. Does the 2016 ALS prevalence report include capture-recapture estimations?** No. It was our hope to publish two prevalence estimates for 2016: one with the established algorithm of actual case counts (lower-bound estimates) and one with capture-recapture approximating the number of missing cases (upper-bound estimates) but this information is not yet publicly available. Analysis of scientific data does not allow for the publication of results without rigorous peer review. A paper outlining the capture-recapture methodology and its results is expected to be published in the *Journal of Neuroepidemiology*. This paper is expected to be published before the end of 2021.
- 10. When will the Registry include the capture-recapture estimates?** We are striving to include the estimated number of missing ALS cases in future reports, starting with the 2017 ALS prevalence report.
- 11. What is the Registry doing to find these missing cases?** We hypothesize that many of the missing cases are from private pay insurance companies (e.g., Health Maintenance Organizations, Preferred Provider Organizations). CDC/ATSDR is reaching out to insurance companies for information and ALS patient organizations (e.g., ALS Association, Muscular Dystrophy Association, Les Turner ALS Foundation) to request data on ALS cases they support through their nationwide offices, chapters, and clinics. This information is cross-referenced with the Registry's data to add any missing cases. The Registry is also cross-referencing its data with the Massachusetts ALS Registry and plans to do so with any other proposed state ALS registries. We are also seeking further approvals to modify the Registry's data collection methods (i.e., allowing new sources of data to be included) in order to allow for improved case counts.
- 12. When will the Registry add additional data from other sources?** The Registry has contacted our partner organizations to begin requesting additional data. Before we are able to receive new data, agreements must be in place. It is our hope that these data agreements will

be active within 12-24 months. These data agreements must be written and finalized to ensure patient privacy and security.

- 13. How will adding missing cases help the Registry?** By adding new cases of ALS to the Registry, we will have a more precise estimate of how many people have ALS in the United States. This will also improve overall case counts in the Registry and strengthen our multi-faceted research platform.
- 14. How will these missing cases help researchers?** Knowing how many persons have ALS is an important way for researchers to measure disease burden (e.g., for patients and caregivers) and to identify where health care improvements such as access to care should be made. In addition, this information assists future clinical trials and research studies on the cause(s) and risk factors of ALS.
- 15. When will the 2017 prevalence report be published?** We have received the 2017 data from CMS and VA and are currently cleaning, removing duplicate information, and analyzing the data. Analysis is anticipated to be completed by the end of 2021. The information will then be submitted for final review and published. CDC/ATSDR recently asked CMS and VA for the 2018 data.