**INTRODUCTION**

- Amyloid light chain (AL) amyloidosis is a rare, progressive, and typically fatal disease caused by extracellular deposition of misfolded immunoglobulin light chains (LCs).
- Soluble toxic aggregates and deposited fibrils (amyloid) lead to progressive failure of vital organs, including the heart and kidneys, causing significant morbidity and mortality.
- There is a lack of recent data on the epidemiology of AL amyloidosis in the United States.
- The objective of this study was to provide an up-to-date estimate of the prevalence and incidence of AL amyloidosis in the United States.

**METHODS**

**Study Design and Data Source**

- Retrospective, cross-sectional study using 2007-2015 Truven MarketScan® commercial and Medicare supplement databases.
- Covering approximately 65 million commercially insured patients and their dependents and 5.3 million Medicare-eligible retired employees.

**Study Population**

- Adults ≥18 years of age with AL amyloidosis if they had 1) incident claim or 2) outpatient claims consistent with AL amyloidosis (International Classification of Diseases, Tenth Revision, Clinical Modification [ICD-10-CM] code E56.4x; E56.8x, or E56.9x) in any medical record within a diagnostic field (ICD-9-CM).
- Underwent 1 AL amyloidosis-specific treatment (eg, chemotherapy, hematopoietic stem cell transplantation) or after the first amyloidosis diagnosis (index date).

**Study Measures**

- Prevalence was calculated as the number of patients with AL amyloidosis divided by the number of enrollees on June 30th of each calendar year and reported as cases per million in each year.
- Incidence was calculated as the number of patients with AL amyloidosis who were disease-free 3 years from diagnosis, continuous enrollment was not required.
- Yearly prevalence proportions were reported for each calendar year from 2007 through 2015.
- Incidence was calculated as the number of patients with AL amyloidosis who were disease-free 3 years from diagnosis.
- APC (annual percentage change) was calculated by fitting a linear regression line to the natural logarithm of the rates.
- There is a lack of recent data on the epidemiology of AL amyloidosis in the United States.

**Statistical Analysis**

- Yearly prevalence proportions were reported for each calendar year from 2007 through 2015.
- Yearly incidence rates were reported from 2008 through 2015.
- Age–gender–adjusted rates to the 2010 US census population were also reported.
- To characterize trends in AL amyloidosis prevalence and incidence rates over time, annual percentage change (APC) was calculated by fitting a linear regression line to the natural logarithm of the ratios using the calendar year as a regressor variable. APC is used to describe rates over time; with this approach, rates are assumed to change at a constant percentage of the previous year's rate.

**RESULTS**

- The overall study included 73,724 prevalent patients with AL amyloidosis (368-1508 unique cases per year) and 2077 incident patients with AL amyloidosis (169-377 cases per year).
- Mean (SD) age for prevalent patients was 66.3 years (12.1); 45% were women, all US regions were represented, and most patients had commercial insurance (Table 1).

**REFERENCES**

1. Rajkumar SV. AL, amyloid light chain; APC, annual percentage change; PMPY, per million person-years.

**DISCUSSION AND CONCLUSIONS**

- This study showed a pattern increased prevalence of AL amyloidosis coupled with stable incidence.
- Although our study could not determine the mechanisms responsible for the observed change, a nationwide Swedish study in 1430 patients with AL amyloidosis diagnosed between 1995 and 2013 found significant improvement in overall survival over time, a change that could explain our findings.
- Extrapolating from our study data, there were 12,000 adults in the US living with AL amyloidosis in 2015, and the number seems likely to continue to rise.
- Limitation – Given the absence of ICD-9-CM or ICD-10-CM codes for AL amyloidosis, we selected codes for the potential of an important clinical endpoint to eliminate as many patients without AL amyloidosis as possible. This requirement would be expected to decrease the sensitivity but to increase the specificity of our approach to detecting the change. However, a patient with transthyretin-related hereditary amyloidosis would still likely have been included in our sample.

**TABLES**

**Table 1. Patient Demographic Characteristics Among Prevalent Cases**

<table>
<thead>
<tr>
<th>Year</th>
<th>N</th>
<th>Age, years (SD)</th>
<th>Male, n (%)</th>
<th>Female, n (%)</th>
<th>South, n (%)</th>
<th>Midwest, n (%)</th>
<th>Northeast, n (%)</th>
<th>West, n (%)</th>
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</table>

**Figure 1. Prevalence of AL amyloidosis in a US commercially insured population, 2007-2015.**

**Figure 2. Incidence of AL amyloidosis in a US population with commercial and Medicare supplement insurance, 2008-2015.**