Use of Commercial Claims Data to Estimate Hereditary Transthyretin-Mediated Amyloidosis Prevalence and Incidence in the US

Duncan Brown, PhD[1]; Montserrat Vera Llonch, MD, MPH, MSc[1]; Jose Tomas Ortiz Perez, PhD[2]; Sheila R. Reddy, PhD, RPh[3]; Eunice Chang, PhD[3]; Marian H Tarbox, MPP[3]; Jose Nativi-Nicolau, MD[4]; Nowell M. Fine, MD, SM[5]

Institutions: [1] Akcea Therapeutics, Boston, MA. [2] Amyloidosis and Myeloma Unit. Hospital Clínic Barcelona, Institut d'Investigacions Biomèdiques August Pi i Sunyer, Universitat de Barcelona. [3] Partnership for Health Analytic Research, LLC, Beverly Hills, CA. [4] School of Medicine, University of Utah, Salt Lake City, UT. [5] University of Calgary, Calgary AB.

BACKGROUND & OBJECTIVE

- Current prevalence and incidence estimates of transthyretin-mediated (ATTR) amyloidosis, including the hereditary subtype (mutant or variable, ATTRv), in the US remain uncertain
  - Estimates are thought to be underestimated due to diagnostic uncertainty caused by a lack of disease awareness and overlapping clinical manifestation[1,5]
- Prevalence of ATTRv amyloidosis has been estimated to be ≤6,400 cases among the US population[2]; however, these estimates are based on small samples and older data[1,3,4]
- The objective of this study was to investigate the feasibility of estimating hereditary transthyretin-mediated (ATTRv) amyloidosis prevalence and incidence, using a large US insurance claims database

METHODS

Retrospective study using IBM® MarketScan® Commercial and Medicare Supplemental databases[a] from 01/01/2014 – 12/31/2018

Patient Identification

- ≥1 inpatient or ≥2 outpatient claims with an ICD-10-CM code for hereditary (E85.1,E85.2) or wild-type (E85.82) amyloidosis form in 2018 or another amyloidosis form in 2018 plus the following between 2014-2018:
  - ≥1 claim for congestive heart failure, cardiomyopathy, or neuropathy; and no chemotherapy, stem cell transplant, or light-chain amyloidosis claims
- Of patients with ATTR identified above, those lacking a code for wild-type (E85.82) were then identified as ATTRv
- Patients with dementia excluded

Study Measures

- 2018 ATTRv incidence rate:
  - Number of cases of newly diagnosed ATTRv in 2018 divided by total at-risk (disease-free) patient years from 01/01/2018 to either diagnosis (cases) or enrollment end (non-cases) in 2018, whichever occurred first
    - At-risk members defined as being ATTRv-free at the beginning of 2018, and who had continuous enrollment, but no ATTR amyloidosis claim (ICD codes: 277.30, 277.31, 277.39; ICD-10-CM: E85.0, E85.1, E85.2, E85.3, E85.4, E85.81, E85.82, E85.89, E85.9) in 2017. Among them, cases were those newly diagnosed with ATTRv in 2018.
  - Incidence reported as cases per million person-years (PMPY)
  - Enrollment was continuous during at-risk period
- 2018 ATTRv prevalence proportion:
  - Number of adult patients who had prevalent (existing or newly diagnosed) ATTRv, divided by all members enrolled in a health plan on June 30th of each 2018
  - Prevalence reported as cases per million persons

Statistical Analysis

- Incidence rates and prevalence estimates stratified by age group and gender
- All data transformations and statistical analyses were performed using SAS© version 9.4

a. MarketScan is a trademark of IBM Corporation in the United States and other countries.

RESULTS

Table 1. ATTRv Amyloidosis Incidence Among Commercially Insured Adults in 2018

	N	Incidence (per million person-years)
All	53	4.46
Age group		
18-34	4	1.23
35-54	14	2.78
55-64	17	5.96
65+	18	24.41
Gender		
Female	21	3.37
Male	32	5.67

ATTRv: hereditary transthyretin.

- ATTRv incidence in 2018 was 4.46 PMPY
  - Incidence was highest among those ≥65 years (24.41) and among males (5.67) vs. females (3.37), all PMPY

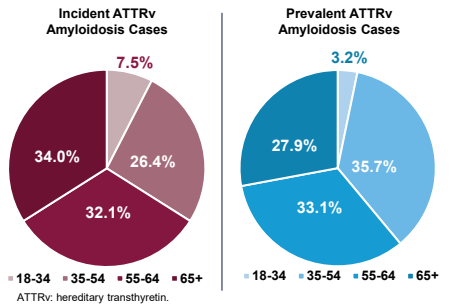
Table 2. ATTRv Amyloidosis Prevalence Among Commercially Insured Adults in 2018

	N	Prevalence (per million persons)
All	154	8.22
Age group		
18-34	5	0.82
35-54	55	7.19
55-64	51	12.85
65+	43	42.18
Gender		
Female	65	6.64
Male	89	9.95

ATTRv: hereditary transthyretin.

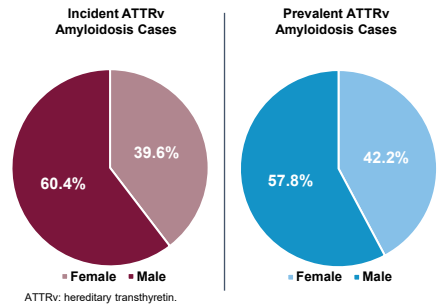
- Estimated prevalence of ATTRv amyloidosis in 2018 of 8.22 per million
  - Prevalence was highest among patients ≥65 years (42.18) and among males (9.95) vs. females (6.64), all per million

Figure 1. Distribution of ATTRv Amyloidosis Cases by Age in 2018



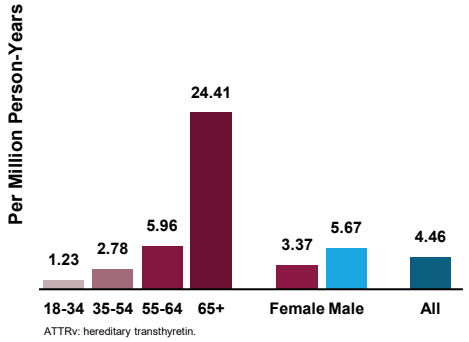
ATTRv: hereditary transthyretin.

Figure 2. Distribution of ATTRv Amyloidosis Cases by Gender in 2018



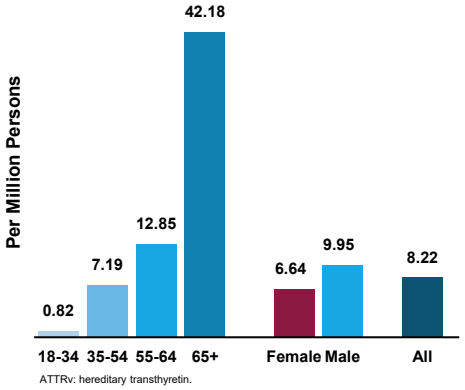
ATTRv: hereditary transthyretin.

Figure 3: ATTRv Amyloidosis Incidence Among Commercially Insured Adults in 2018



ATTRv: hereditary transthyretin.

Figure 4: ATTRv Amyloidosis Prevalence Among Commercially Insured Adults in 2018



ATTRv: hereditary transthyretin.

LIMITATIONS

- Estimation of ATTRv incidence and prevalence using administrative claims data has not been previously validated; such estimation is difficult due to diagnostic challenges such as lack of awareness of the disease and, until recently, the absence of medical coding specific to the different types of transthyretin amyloidosis, including ATTRv and wild-type ATTR amyloidosis (ATTRwt)
- Estimates in this study are specific to commercially insured adults in the US and are not generalizable to the broader US adult population
  - Further, we determined that our ATTR prevalence estimates were lower compared to age-standardized estimates for the US population

CONCLUSIONS

- This study developed a commercial claims-based algorithm to estimate prevalence and incidence of ATTRv in the US
- This algorithm may improve understanding of ATTRv epidemiology

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dbrown@akceatx.com

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DISCLOSURES

- SRR, EC, and MHT are employees of Partnership for Health Analytic Research, LLC, which was paid by Akcea to perform this research.
- JP: Advisory board fees: Akcea
- JN: Financial: Pfizer, Akcea and Eidos; Grants: Pfizer. Consultant: Pfizer, Eidos, Akcea, and Alnylam.
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