Epidemiology of transthyretin amyloid cardiomyopathy (ATTR-CM) in France, a study based on the systeme national des donnees de sante (SNDS) the French nationwide claims database

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Transthyretin (TTR) Amyloid Cardiomyopathy (ATTR-CM) is a rare, progressively debilitating, fatal disease with poor prognosis caused by amyloid deposition of fibrils derived from the serum protein TTR in the extracellular matrix of the heart. As amyloid infiltration in the heart progresses, atrial and ventricular walls thicken and become restrictive, resulting in diastolic dysfunction and further progression leads to heart failure, usually with preserved ejection fraction. Systolic dysfunction occurs in the late stages of the disease. Cardiac symptoms and conduction abnormalities also become increasingly clinically apparent (e.g. fatigue, shortness of breath, syncope or arrhythmias) with progression over time.

Epidemiology of ATTR-CM is poorly understood as there are few existing studies that estimate its frequency in the general population. We aimed to estimate the prevalence and the incidence of ATTR-CM in France between 2011 and 2017, to describe demographic characteristics of incident cases and to assess patient survival.

We used data from the SNDS database, which collects all national health insurance and hospital discharge data. As there is no specific ICD-10 marker code for ATTR-CM used in the SNDS, ATTR-CM diagnosis required both an amyloidosis and a cardiovascular condition, not necessarily reported at the same visit. Diagnostic date was defined when the features from amyloidosis and cardiovascular conditions were selected as events. Patients with a probable AL form of the disease were excluded. To remain conservative, patients younger than 50 yo were also excluded.

Between 2011 and 2017, 4,815 patients with incident ATTR-CM were identified. Incidence rate was multiplied by more than 3 times, from 0.5 / 100,000 person-year in 2011 to 1.8 / 100,000 person-year in 2017, reaching 1,225 new cases in 2017. Sex-ratio remained stable (2:1). Most of the 4,815 identified patients were older than 70 yo. In the group ATTR CM >70 yo, there were 3 times more men than women. Median age at diagnosis was 84.0 for women and 82.0 for men. The range of 80-89 yo represents about 50% of diagnose rate in overall population.

Median survival was 33.7 months overall with minor differences between gender. Survival probability was 0.69 a year after diagnosis, 0.58 two years after, 0.48 three years after, and 0.41 four years after. Using exponential modeling the life expectancy of this population would be about 4 years for the 20 next years.

This study is, to our knowledge, the first estimate of ATTR-CM incidence rates based upon an analysis of a national claims database. Our findings are consistent with existing data concerning the frequency of amyloidosis, even though were only identified the ICD-10 coded diagnosed cases, leading to possible underestimation of the full prevalence of the disease in France. SNDS demonstrated to be a scientifically valid data base to follow-up the standard of care of ATTR-CM.