Patients' description and resource use in Wilson disease patients based on French and Japanese claims real-world databases

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Context & objectives

Wilson disease (WD) is a rare and inherited disorder of copper metabolism caused by mutations in the copper transporter ATP7B gene, which leads to toxic hepatic and brain accumulation of copper. Symptoms (hepatic, neurological and/or psychiatric) are very variable and most frequently first develop in adolescence or early adulthood. Left untreated, the condition progresses to severely debilitating complications and death.

Medical treatment currently relies on copper chelators (D-penicillamine or trientine) or zinc salts. Poor treatment adherence is frequent and may result in severe complications.

The main objective of the study was to describe WD patient's characteristics in France and Japan. The secondary objectives were to describe disease management and burden of WD and to estimate adherence to treatment.

Methods

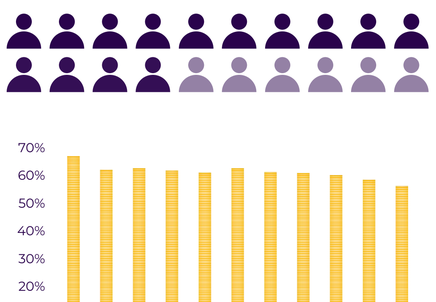
This retrospective observational study analysed patients with WD in the SNDS (France) and in MDV (Japan) databases, using ICD10 code E830; the study included French patients identified between 2009 and 2019, and followed until 2019, and Japanese patients identified between 2011 and 2021, and followed until 2021.
Treatments were identified through CIP codes in France and receipt codes in Japan. Symptoms (neurological, hepatological or psychiatric) were tracked using ICD10 and WHO ATC codes in France, disease and receipt codes in Japan.
Deaths were extracted (all registered deaths in France, deaths in hospital in Japan).

Patients' description

Mean age: 40 (France) | 55 (Japan)

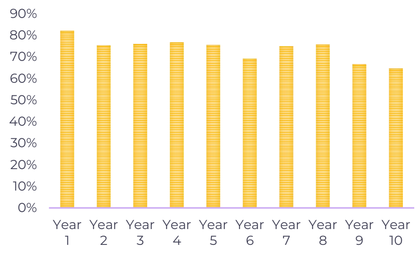
% of males: 52% (France) | 55% (Japan)

Presence of symptoms



% at inclusion: 82% (France) | 82% (Japan)

% during follow-up

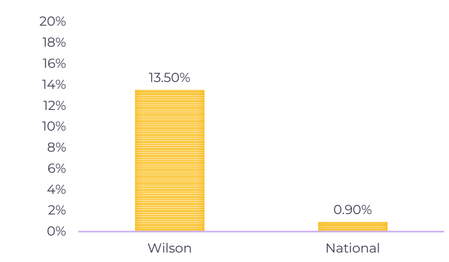


WD treatment

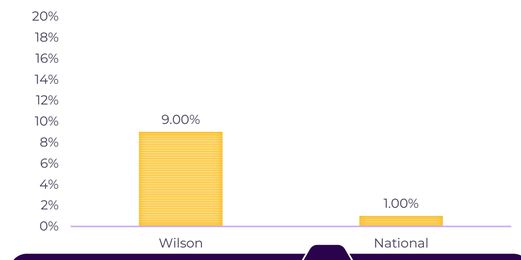
% with at least one treatment during follow-up

Median treatment duration: 0 day (France) | 42 days (Japan)

Mortality



Rate



Mean age at death with WD: 59 (France) | 69 (Japan)

National life expectancy: 80 86 (France) | 82 88 (Japan)

Conclusion

These results indicate that WD management may be improved in both countries, since a high proportion of patients experienced symptoms despite treatment. Survival was lower in each of the WD populations than in the corresponding general populations. These conclusions are similar in France and Japan, although in Japan, a higher proportion of patients received a WD-specific treatment and age of death was higher.
Conducting the same study on databases from different countries helps to compare disease care management, showing different drug prescription patterns and better observance in the Japanese population.