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ABSTRACT

An Epidemiological Study of Wilson's Disease in Morocco.

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Objectives: Wilson's disease is an autosomal recessive disorder that affects copper metabolism leading to copper accumulation in the liver, nervous system, and cornea. There is a lack of data regarding the epidemiological, clinical, laboratory, treatment and survival from Wilson's disease cohorts in Morocco. The aim of this study is to examine these features and the cause of death in a Moroccan population

Methods: The study was carried out at the University Hospital Center of Fez, Morocco; 60 patients were diagnosed with Wilson's disease from 2008 to 2020. The diagnosis was based on low serum ceruloplasmin, increased urinary copper concentrations, the presence of Kayser–Fleischer rings, family history of Wilson's disease and a Leipzig score ≥ 4 .

Results: Fifty-two patients were referred for a hepatic or neurological pathology; eight patients were asymptomatic. Consanguinity of marriage was found in 56.7% of cases. The mean duration of illness (52 patients) was 9.3 ± 9 months. Kayser–Fleischer rings were found in 62.3% of 53 patients. 41/43 (95.3%) patient had low serum ceruloplasmin ($< 0.2\text{g/L}$) and 24-hours urinary copper $> 100 \mu\text{g/day}$ was found in all cases. The treatment was established with D-penicillamine in 54 patients, zinc acetate in one patient, zinc sulphate in one patient and four patients no treated. D-penicillamine was discontinued in 12 patients because of adverse effects such nausea, severe vomiting and diarrhea, neurological deterioration, proteinuria and toxiderma. Forty-seven patients were clinically and biologically stabilized, and 13 patients died (21.7%). The main cause of death was the diagnosis in advanced stage of disease and stopping treatment.



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Conclusions: Wilson's disease is one of the rare diseases treated efficacy, but the late diagnosis and stopping treatment can cause a high mortality.

Keywords: Wilson's disease, epidemiology, mortality, Fez, Morocco.

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