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Controversies and advances in Wilson's disease (WD) 2017

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Wilson's disease is a rare inherited disorder with variable epidemiology. Geographical areas with high rates consanguinity like Costa Rica and Sardinia, Italy show the highest prevalence of hepatolenticular degeneration. Costa Rica reports the highest incidence worldwide 4.9/100.000, while in some areas it can reach more than 50.0/100.000. As a consequence, since 1980 efforts have been established to promote early diagnosis, family screening and management (Figure 1). Diagnosis is based on two abnormal levels of cooper and Leipzig criteria. Costa Rican genetic mutations are mainly Asn1270Ser the same mutation found in Sicilian (Italy) WD patients. Due to the high incidence of WD in Costa Rica, some aspects are still needed to optimize management such as:

Early Detection of Asymptomatic Patients: In high incidence areas, the detection must be implemented by screening family members for every confirmed patient; through ceruloplasmin determination and 24h urinary copper excretion in patients having low or normal-low ceruloplasmin level.

Excellent Laboratory: All tests and copper determinations (i.e ceruloplasmin, 24h urinary copper excretion, serum free copper levels, genetic mutations, hepatic copper) must be done in a specific centralized laboratory.

Fulminant Hepatic Failure Secondary to WD: Fulminant WD constitutes the worst clinical presentation of WD in which diagnosis is frequently late. FWD patients are generally women between 10-20 years old, with acute liver failure, hemolysis and liver transplant candidates. When transplant is not an option, treatment is done with prostaglandins, vitamin E and haemoperfusion administration. In Costa Rica, 4 FWD patients who didn't undergo transplant survived using prostaglandins and vitamin E. Although the etiology of FWD is mainly unknown, one case of viral diarrhea before the manifestation was reported.

Neurological Manifestations: Early diagnosis and structured physical rehabilitation programs are essential for adequate management.

New Treatments: It is crucial to allocate resources in research to develop and evaluate new drugs (curcuma, trientene nanomolecules, tetrathiomolybdate, genetic treatment) to improve diagnostic and management options.

Recent Publications:

1. Hevia et al (2017) National Alliance for Wilson´s disease: health policy in Costa Rica. Hepatology, Medicine and Policy 2:5
2. Rodriguez, Hevia and Sturniolo (2015) Wilson´s disease: A review of what we have learned. World Journal of Hepatology 7 (29): 2859-2870
3. Hevia F and Miranda M (1989) The special problem of Wilson´s disease in Costa Rica. An unexpected high prevalence. Gastroenterol. Int 22:228
4. Hevia F J (2009) Practical management of Wilson´s disease. International Hepatology Updates 63-79

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