When is the suitable time to perform follow-up liver biopsies in Wilson disease patients?

April 8 2010

Wilson disease is a rare autosomal recessive disorder of copper accumulation that is characterized by hepatic, neurological and psychiatric manifestations. USA researchers recently investigated the progression of hepatic histopathology in serial liver biopsies from Wilson disease patients and estimated that liver biopsy with hepatic copper quantification every 3 years should be considered.

As a result of the rarity of Wilson disease (WD) and the fact that liver biopsy is not performed routinely during follow-up, unless clinically indicated, the progression and timing of the liver pathology and its correlation with different anti-copper treatments or aminotransferase levels are poorly characterized. Previous studies have demonstrated the possibility of improvement of steatosis and inflammation grade, and of fibrosis stage during long-term follow-up. However, studies on serial liver biopsies, as well as studies on the correlation between hepatic histology and clinical parameters, are lacking.

A research article to be published on March 28 , 2010 in the World Journal of Gastroenterology addresses this question. A study conducted by the University of California Davis (USA), Texas Children's Hospital (USA), and University of Padua (Italy) under the guidance of Dr. Valentina Medici studied the evolution of liver histology in WD patients during penicillamine and zinc treatment, to define the rate of progression of liver damage and to correlate the clinical and biochemical parameters of liver injury with hepatic copper concentration.

The estimated rate of progression of hepatic fibrosis (as result of the mean difference in fibrosis scores divided by the mean interval in years between the first and second liver biopsies) in the entire group was 0 units per year between the first and second liver biopsy, and 0.25 between the second and third. However, among progressors, the rate of progression of fibrosis was estimated at 0.23 fibrosis units per year between the first and second biopsy, and 0.6 units between the second and third. Progressors showed a mean hepatic copper concentration higher than non-progressors at all time points. The histological progression did not correlate with subsequent aminotransferase levels or with type of therapy.

The observation of the inability of clinical tools to detect the progression of fibrosis despite treatment suggests that a liver biopsy with hepatic copper quantification every 3 years should be considered.

More information: Cope-Yokoyama S, Finegold MJ, Sturniolo GC, Kim K, Mescoli C, Rugge M, Medici V. Wilson disease: Histopathological correlations with treatment on follow-up liver biopsies. World J Gastroenterol 2010; 16(12): 1487-1494 www.wjgnet.com/1007-9327/full/v16/i12/1487.htm

Provided by World Journal of Gastroenterology