

The authors' aim in this paper is to assess the use of biotinidase as a predictor of hepatic synthetic function concentrations in pediatric patients with chronic liver disease. Plasma biotinidase activities were statistically significantly lower in children with decompensated liver disease compared to children in the control group. The authors found no statistically significant differences between children with compensated liver disease and those in the control group. There were statistically significant positive correlations between plasma biotinidase activities and albumin and prothrombin concentrations and significantly positive inverse correlations with prothrombin times and INR. The authors suggest that plasma biotinidase concentrations can be used as a predictor for hepatic synthetic function impairment. They recommended that patients with decompensated liver disease be supplemented with biotin to protect them from biotin deficiency because of lower biotinidase activity.

Comments:

The actual study was performed with only 13 of the patients with decompensated liver disease and had a variety of different diagnoses. All these decompensated patients are placed in a single group regardless of whether the liver disease associated with this disorder is actually cirrhosis, hepatitis, etc. Only the decompensated patients with cirrhosis should have been included!

The authors should have indicated the actual biotinidase activity for each patient with their specific diagnosis to determine if those with more "normal" activities actually do not have hepatic synthetic dysfunction.

The correlations between the various studied parameters should not include individuals with compensated liver disease. In fact, if you remove the patients with non-cirrhotic liver disease the correlations would likely be stronger! However, the sample size would be too small for a good study.

The authors have only confirmed what other studies have previously reported (References 2, 10, 12, 13). They omitted the first report demonstrating the positive correlation of biotinidase activity with albumin concentration in patients with cirrhotic liver disease.

- Grier RE, Heard GS, Watkins P, Wolf B. Low biotinidase activities in the sera of patients with impaired liver function: evidence that the liver is the source of serum biotinidase. [Clin Chim Acta](#). 1990;186:397-400.

The only new thing in this paper is the authors' suggestion that patients with decompensated liver disease who have lower biotinidase activity be supplemented with biotin. This recommendation is not founded on fact! The lowest biotinidase activity that was reported in their decompensated group was just below an

activity of 4. This activity would be about the same activity of an individual who is a heterozygote for profound biotinidase deficiency that is the activity of a parent of a patient with profound biotinidase activity. These heterozygotes are not symptomatic with this degree of activity and are not supplemented with biotin. Therefore, although biotin supplementation would not be harmful, there is no reason to supplement any of the decompensated patients with biotinidase activities in the ranges reported in this paper! Therefore, why supplement the patients in their population.

This study does not really add to what has already been reported. In addition, the authors present the data on a small group of patients with a varied group of diseases and these data are not appropriately analyzed, nor bases on the sample sizes can it be appropriately analyzed. The authors conclusions are similar to others, but with incomplete data and their basic recommendation is not founded.