## Abstract

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## Urinary excretion of 3-hydroxyisovaleryl carnitine is an early and sensitive indicator of marginal biotin deficiency in humans.

Stratton SL, Horvath TD, Bogusiewicz A, Matthews NI, Henrich CL, Spencer HJ, Moran JH, Mock DM.

Department of Biochemistry and Molecular Biology, College of Medicine, University of Arkansas for Medical Sciences, Little Rock, AR 72205.

**OBJECTIVE**: Mounting evidence indicates that marginal biotin deficiency is not rare, contrary to previous assumptions. Accordingly, robust indicators of biotin status would be useful.

**METHODS**: In a study of 10 healthy adults, we recently provided evidence that abnormally increased plasma concentration of 3-hydroxyisovaleryl carnitine (3HIA-carnitine) is a sensitive indicator of marginal biotin deficiency. We sought to determine whether urinary excretion of 3HIA-carnitine (expressed as the ratio to urinary creatinine) significantly increases in marginal biotin deficiency. Marginal, asymptomatic biotin deficiency was induced experimentally in the same 10 healthy adults (8 women) by feeding undenatured egg white with meals for 28 d. Biotin status was repleted by a mixed general diet plus biotin supplementation. Urinary excretion of 3HIA-carnitine was determined by liquid chromatography-tandem MS on d 0, 14, and 28 (depletion) and on d 35 and 50 (repletion).

**RESULTS**: Mean urinary 3HIA-carnitine concentration increased with depletion (P < 0.0001; d 0 vs. 28) and decreased with repletion (P = 0.0002; d 28 vs. 50). Urinary 3HIA-carnitine excretion was greater than the upper limit of normal in 9 of 10 participants by d 14 and decreased to within normal limits by d 50 in all participants.

**CONCLUSION**: This study provides evidence that urinary excretion of 3HIA-carnitine is an early and sensitive indicator of marginal biotin deficiency. The ease of collection of untimed urine samples and application of a new analytical method with simplified sample preparation suggest that urinary 3HIA-carnitine is likely to be a useful indicator for large population studies.

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