## 3-Hydroxyisovalerylcarnitine

Cat. No.: HY-N11200 CAS No.: 99159-87-2 Molecular Formula:  $C_{12}H_{23}NO_5$ Molecular Weight: 261.31

Target: **Endogenous Metabolite** Pathway: Metabolic Enzyme/Protease Storage: -20°C, protect from light

\* In solvent: -80°C, 6 months; -20°C, 1 month (protect from light)

**Product** Data Sheet

## **BIOLOGICAL ACTIVITY**

Description

3-Hydroxyisovalerylcarnitine is a carnitine derivative. 3-Hydroxyisovaleryl carnitine in plasma may serve as a novel biomarker of biotin deficiency in humans, with its concentration increasing with biotin deficiency. Biotin deficiency reduces the activity of biotin-dependent 3-methylcrotonyl-CoA carboxylase, hinders the conversion of 3-methylcrotonyl-CoA into 3methylglutaconyl-CoA, and impairs the leucine catabolism pathway; resulting in plasma 3-Hydroxyisovalerylcarnitine concentrations rise<sup>[1]</sup>.

## **REFERENCES**

[1]. Horvath TD, et al. Quantitative measurement of plasma 3-hydroxyisovaleryl carnitine by LC-MS/MS as a novel biomarker of biotin status in humans. Anal Chem. 2010 May 15;82(10):4140-4.

Caution: Product has not been fully validated for medical applications. For research use only.

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