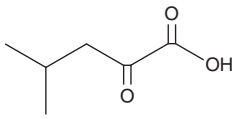
# PRODUCT INFORMATION

## α-Ketoisocaproic Acid

*Item No. 34749*

**CAS Registry No.:** 816-66-0
**Formal Name:** 4-methyl-2-oxo-pentanoic acid
**Synonyms:** 2-Ketoisocaproate, α-Ketoisocaproate, 2-Ketoisocapronic Acid, KIC, 4-MOP, 2-Oxoisocaproic Acid, 4-methyl-2-Oxopentanoate, 4-methyl-2-Oxovaleric Acid, 4-methyl-2-Oxovalerate
**MF:** $C_6H_{10}O_3$
**FW:** 130.1
**Purity:** ≥95%
**UV/Vis.:** $\lambda_{max}$: 241 nm
**Supplied as:** A liquid
**Storage:** -20°C
**Stability:** ≥2 years




*Information represents the product specifications. Batch specific analytical results are provided on each certificate of analysis.*

### Laboratory Procedures

α-Ketoisocaproic acid is supplied as a liquid. A stock solution may be made by dissolving the α-ketoisocaproic acid in the solvent of choice, which should be purged with an inert gas. α-Ketoisocaproic acid is soluble in organic solvents such as ethanol, DMSO, and dimethyl formamide. The solubility of α-ketoisocaproic acid in these solvents is approximately 30 mg/ml.

### Description

α-Ketoisocaproic acid is a metabolite of L-leucine (Item No. 34342) catabolism.[1] It increases the production of reactive species and decreases the activity of mitochondrial complex I, also known as NADH dehydrogenase, and complex II-III in the rat hippocampus when administered at an intracerebroventricular dose of 4 µmol.[2] Urine levels of α-ketoisocaproic acid are reduced in *db/db* diabetic mice compared with heterozygous *db/m* non-diabetic mice.[3] α-Ketoisocaproic acid accumulates in the tissues and body fluids of patients with maple syrup urine disease, an inborn error of metabolism characterized by branched-chain α-keto acid dehydrogenase (BCKAD) deficiency and leads to progressive ketoacidosis, failure to thrive, neurological dysfunction, and, potentially, death.[2]

### References


1. Schiff, M., Ogier de Baulny, H., and Dionisi-Vici, C. Branched-chain Organic Acidurias/Acidaemias. *Inborn Metabolic Diseases. Diagnosis and Treatment*. Saudubray, J.-M, Baumgartner, M.R., and Walter, J., editors, 6th edition, *Springer* (2016).
2. Farias, H.R., Gabriel, J.R., Cecconi, M.L., *et al.* The metabolic effect of α-ketoisocaproic acid: *In vivo* and *in vitro* studies. *Metab. Brain Dis.* **36(1)**, 185-192 (2021).
3. Kim, N.H., Hyeon, J.S., Kim, N.H., *et al.* Metabolic changes in urine and serum during progression of diabetic kidney disease in a mouse model. *Arch. Biochem. Biophys.* **646**, 90-97 (2018).





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