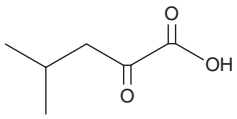
PRODUCT INFORMATION

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.: λ_{max}: 241 nm
Supplied as: A liquid
Storage: -20°C
Stability: ≥4 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).

WARNING
THIS PRODUCT IS FOR RESEARCH ONLY - NOT FOR HUMAN OR VETERINARY DIAGNOSTIC OR THERAPEUTIC USE.

SAFETY DATA
This material should be considered hazardous until further information becomes available. Do not ingest, inhale, get in eyes, on skin, or on clothing. Wash thoroughly after handling. Before use, the user must review the complete Safety Data Sheet, which has been sent *via* email to your institution.

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