

PRODUCT INFORMATION



Isovaleryl-Coenzyme A (sodium salt)

Item No. 39999

Formal Name: S-(3-methylbutanoate) coenzyme A, sodium salt

Synonym: Isovaleryl-CoA

MF: C₂₆H₄₄N₇O₁₇P₃S • XNa

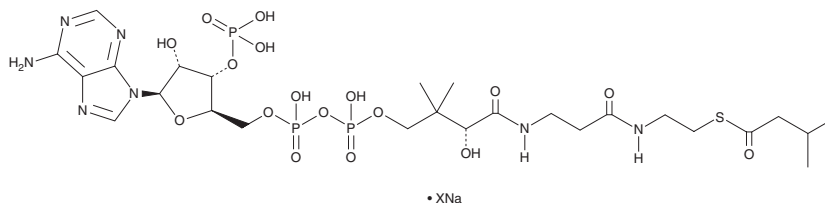
FW: 851.7

Purity: ≥95%

Supplied as: A solid

Storage: -20°C

Stability: ≥4 years



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

Laboratory Procedures

Isovaleryl-coenzyme A (isovaleryl-CoA) (sodium salt) is supplied as a solid. A stock solution may be made by dissolving the isovaleryl-CoA (sodium salt) in water. We do not recommend storing the aqueous solution for more than one day.

Description

Isovaleryl-CoA is a branched-chain acyl CoA and an intermediate in the metabolism of the essential amino acid L-leucine (Item No. 34342).^{1,2} It is formed from L-leucine by the branched-chain α -keto acid dehydrogenase (BCKAD) complex and is converted to 3-methylcrotonyl-CoA by isovaleryl-CoA dehydrogenase (IVD), an enzyme involved in the inborn error of metabolism isovaleric acidemia.³ Isovaleryl-CoA increases the oxidation rate of the branched-chain 2-oxo acids 3-methyl-2-butanoate and 4-methyl-2-oxopentanoate in isolated rat muscle mitochondria in the absence of carnitine when used at a concentration of 1 mM.⁴ It also inhibits succinate-CoA ligase activity in rat liver mitochondria (IC₅₀ = 273 μ M).⁵

References

1. Duan, Y., Li, F., Li, Y., *et al.* The role of leucine and its metabolites in protein and energy metabolism. *Amino Acids* **48(1)**, 41-51 (2016).
2. Mohsen, A.-W.A., Anderson, B.D., Volchenboum, S.L., *et al.* Characterization of molecular defects in isovaleryl-CoA dehydrogenase in patients with isovaleric acidemia. *Biochemistry* **37(28)**, 10325-10335 (1998).
3. Mack, M., Schniegler-Mattox, U., Peters, V., *et al.* Biochemical characterization of human 3-methylglutaconyl-CoA hydratase and its role in leucine metabolism. *FEBS J.* **273(9)**, 2012-2022 (2006).
4. Veerkamp, J.H., van Moerkerk, H.T.B., and Wagenmakers, A.J.M. Interaction of short-chain and branched-chain fatty acids and their carnitine and CoA esters and of various metabolites and agents with branched-chain 2-oxo acid oxidation in rat muscle and liver mitochondria. *Int. J. Biochem.* **17(9)**, 967-974 (1985).
5. Bergen, B.J., Stumpf, D.A., Haas, R., *et al.* A mechanism of toxicity of isovaleric acid in rat liver mitochondria. *Biochem. Med.* **27(2)**, 154-160 (1982).

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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