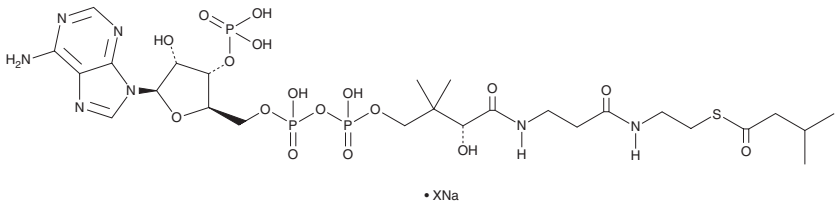
PRODUCT INFORMATION

Isovaleryl-Coenzyme A (sodium salt)

Item No. 39999

Formal Name:	S-(3-methylbutanoate) coenzyme A, sodium salt
Synonym:	Isovaleryl-CoA
MF:	$C_{26}H_{44}N_7O_{17}P_3S \bullet XNa$
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 (IC_{50} = 273 µM).[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).
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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 branced-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).

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