

## Produktinformation



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# **PRODUCT** INFORMATION



## Isovaleryl-L-carnitine-d<sub>3</sub> (chloride)

Item No. 26574

Formal Name:	(R)-3-carboxy-N,N-dimethyl-N-(methyl-d <sub>3</sub> )-2-((3-methylbutanoyl)oxy)propan-1-aminium, monochloride	1
Synonym:	L-Isovalerylcarnitine-d <sub>3</sub>	
MF:	$C_{12}H_{21}D_3NO_4 \bullet CI$	D N+
FW:	284.8	
Chemical Purity:	≥98% (IsovaleryI-L-carnitine)	
Deuterium		
Incorporation:	≥99% deuterated forms (d <sub>1</sub> -d <sub>3</sub> ); ≤1% d <sub>0</sub>	
Supplied as:	A solid	O OH
Storage:	-20°C	
Stability:	≥2 years	

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

#### Laboratory Procedures

IsovaleryI-L-carnitine-d<sub>3</sub> (chloride) is intended for use as an internal standard for the quantification of isovaleryl-L-carnitine (chloride) (Item No. 26555) by GC- or LC-MS. The accuracy of the sample weight in this vial is between 5% over and 2% under the amount shown on the vial. If better precision is required. the deuterated standard should be quantitated against a more precisely weighed unlabeled standard by constructing a standard curve of peak intensity ratios (deuterated versus unlabeled).

Isovaleryl-L-carnitine- $d_3$  (chloride) is supplied as a solid. A stock solution may be made by dissolving the isovaleryl-L-carnitine-d<sub>3</sub> (chloride) in the solvent of choice. Isovaleryl-L-carnitine-d<sub>3</sub> (chloride) is soluble in organic solvents such as ethanol, DMSO, and dimethyl formamide (DMF), which should be purged with an inert gas. The solubility of isovaleryl-L-carnitine-d<sub>3</sub> (chloride) in ethanol is approximately 25 mg/ml and approximately 20 mg/ml in DMSO and DMF.

#### Description

IsovaleryI-L-carnitine is a naturally occurring acylcarnitine that is formed via metabolism of L-leucine.<sup>1</sup> It increases survival and decreases apoptosis in hepatocyte growth factor-deprived murine C2.8 hepatocytes when used at a concentration of 1 mM.<sup>2</sup> IsovaleryI-L-carnitine inhibits amino acid deprivation-induced proteolysis and autophagy in isolated perfused rat liver when used at concentrations of 77 and 100 µM, respectively.<sup>3</sup> Increased levels of isovaleryl carnitine are associated with isovaleryl-CoA dehydrogenase deficiency (isovaleric acidemia).<sup>4</sup>

#### References

- 1. Cavazza, C. L-carnitine derivatives as therapeutical agents for treating myopathies and neuronal degeneration and for inhibiting proteolysis. Sigma-Tau Industrie Farmaceutiche Riunite S.p.A. US5227518A (1993).
- 2. Revoltella, R.P., Dal Canto, B., Caracciolo, L., et al. L-carnitine and some of its analogs delay the onset of apoptotic cell death initiated in murine C2.8 hepatocytic cells after hepatocyte growth factor deprivation. Biochim. Biophys. Acta 1224(3), 333-341 (1994).
- 3. Miotto, G., Venerando, R., Khurana, K.K., et al. Control of hepatic proteolysis by leucine and isovaleryl-L-carnitine through a common locus. Evidence for a possible mechanism of recognition at the plasma membrane. J. Biol. Chem. 267(31), 22066-22072 (1992).
- 4. Rinaldo, P., Cowan, T.M., and Matern, D. Acylcarnitine profile analysis. Genet. Med. 10(2), 151-156 (2008).

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

#### SAFFTY DATA

al 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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